The Development and Evaluation of a Psycho-education Booklet for Genetic Aortic Disorders

Joycelyn Ling

Supervisors:
Dr. Ilona Juraskova
Prof Richmond Jeremy

This thesis is submitted to The University of Sydney in fulfilment of the requirement for the double degree of Doctor of Clinical Psychology/Master of Science, 2016.
Statement of Originality

This is to certify that to the best of my knowledge, the content of this thesis is my own work. This thesis has not been submitted for any degree or other purposes.

I certify that the intellectual content of this thesis is the product of my own work and that all the assistance received in preparing this thesis and sources have been acknowledged.

Signature: [Signature]

Joycelyn Ling
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To my husband, this is the last thesis, I promise.

To my baby boy who came into this world around Chapter 4, this is dedicated to you.
# Table of Contents

Acknowledgements .................................................................................................................. 3
Table of Contents ...................................................................................................................... 4
List of Tables ............................................................................................................................ 7
List of Figures ........................................................................................................................... 8
Glossary of Terms ..................................................................................................................... 9
Abstract .................................................................................................................................. 10

Chapter 1: Introduction ........................................................................................................... 12
1.1. Genetic Aortic Disorders ................................................................................................. 12
   1.1.1. Marfan Syndrome (MFS) ....................................................................................... 16
   1.1.2. Loey-Dietz Syndrome (LDS) ............................................................................... 18
   1.1.3. Thoracic Aortic Aneurysm and Dissection (TAAD) ............................................... 19
   1.1.4. Bicuspid Aortic Valve Aneurysm Syndrome (BAVAS) ........................................... 19
1.2. Psychosocial Impact of GA Disorders ........................................................................... 20
   1.2.1. Illness Uncertainty ............................................................................................... 21
   1.2.2. Psychological Impact ........................................................................................... 23
   1.2.3. Social Impact ....................................................................................................... 24
   1.2.4. Vocational Impact ............................................................................................... 25
   1.2.5. Health-Related Quality of Life ............................................................................. 26
1.3. Adherence to treatment in Chronic Illness ..................................................................... 27
1.4. Coping and Adjustment to a Diagnosis of GA Disorder .................................................. 29

Chapter 2: Patient Information Needs ..................................................................................... 36
2.1 Information Needs of Individuals with Chronic Conditions ............................................. 36
   2.1.1 Patient Health Education ....................................................................................... 36
   2.1.2 Health literacy and Patient Knowledge of their Condition ....................................... 38
   2.1.3 What patients want to know about their medical condition ................................ ..... 42
   2.1.4 Health Information Sources: Online versus Printed Materials ................................ 46
2.2 Written Health Information ............................................................................................... 49
2.2.1 Purpose and Objectives of Health Information ................................................. 49
2.2.2 Developing Written Health Information .......................................................... 51
2.3 Evaluation of Written Health Information ........................................................ 59
  2.3.1 Outcomes of Evaluation Studies ..................................................................... 59
  2.3.2 Tools and Guidelines for the Evaluation of Written Materials ................. 64
2.4 Aims of the Research ......................................................................................... 68

Chapter 3: The Development of a Psycho-Education Booklet for Individuals with a Genetic Aortic Disorder ................................................................................. 69
3.1 Development and Evaluation of the Booklet ................................................. 69
  3.1.1 Overall process – Introduction ..................................................................... 69
  3.1.1.1 Assess need ............................................................................................. 71
  3.1.1.2 Assess feasibility ..................................................................................... 71
  3.1.1.3 Define the objectives of the booklet ..................................................... 71
  3.1.1.4 Select the design and measures to evaluate the booklet .................... 73
  3.1.1.5 Plan dissemination ................................................................................ 73
3.2 The development of the booklet ...................................................................... 74
3.3 Rigour and quality of the booklet .................................................................... 83

Chapter 4: Pilot Evaluation of the Booklet ......................................................... 87
4.1 Aims .................................................................................................................. 87
4.2 Design ............................................................................................................. 87
4.3 Participants ....................................................................................................... 88
4.4 Procedure ......................................................................................................... 89
  4.4.1 Ethics .......................................................................................................... 89
  4.4.2 Procedure - Patient Group ......................................................................... 89
  4.4.3 Procedure - Clinician group ....................................................................... 91
4.4.4 Quantitative Measures ................................................................................. 91
4.4.5 Qualitative Measures ................................................................................... 96
4.4.6 Data Analysis ............................................................................................... 96
4.5 Results ............................................................................................................. 98
List of Tables

Chapter 3
Table 3.1 Booklet Content List

Chapter 4
Table 4.1 DASS21 scores and corresponding descriptors
Table 4.2 Demographic characteristics of the patient group
Table 4.3 Clinical characteristics of the patient group
Table 4.4 Information sources accessed
Table 4.5 Psychological well-being profile
Table 4.6 Feedback about the content of the booklet (frequency of responses)
Table 4.7 Feedback about the format of the booklet (frequency of responses)
Table 4.8 Feedback about the acceptability of the booklet (frequency of responses)
Table 4.9 Demographic and professional characteristics of the clinician group
Table 4.10 Feedback about the acceptability of the booklet (frequency of responses)
Table 4.11 Feedback about the content of the booklet (frequency of responses)
Table 4.12 Feedback about the format of the booklet (frequency of responses)
List of Figures

Chapter 1

Figure 1.1 Integrated model of adjustment in GA disorders (Connors et al., 2015)

Chapter 3

Figure 3.1 Booklet -Front Page
Figure 3.2 Booklet -Patient’s Story
Figure 3.3 Booklet –What is an Aortic Aneurysm?
Figure 3.4 Booklet –Myths and Misconceptions
Figure 3.5 Booklet –Questions to Ask Your Physician
## Glossary of Terms

<table>
<thead>
<tr>
<th>Abbreviation</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>BAVAS</td>
<td>Bicuspid Aortic Valve Aneurysm Syndrome</td>
</tr>
<tr>
<td>CPS</td>
<td>Control Preferences Scale</td>
</tr>
<tr>
<td>DASS21</td>
<td>Depression, Anxiety and Stress Scale</td>
</tr>
<tr>
<td>DSES</td>
<td>Decision Self-Efficacy Scale</td>
</tr>
<tr>
<td>FBN1</td>
<td>Fibrillin-1 gene</td>
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<tr>
<td>GA Disorders</td>
<td>Genetic aortic disorders</td>
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<tr>
<td>IPDASi</td>
<td>International Patient Decision Aid Standards instrument</td>
</tr>
<tr>
<td>ISQ</td>
<td>Information Style Questionnaire</td>
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<tr>
<td>LDS</td>
<td>Loeys-Dietz syndrome</td>
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<tr>
<td>MFS</td>
<td>Marfan syndrome</td>
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<tr>
<td>NIAMS</td>
<td>National Institute of Arthritis and Musculoskeletal and Skin Diseases</td>
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<tr>
<td>RPAH</td>
<td>Royal Prince Alfred Hospital</td>
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<tr>
<td>TAAD</td>
<td>Thoracic Aortic Aneurysm and Dissection</td>
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Abstract

Introduction: Genetic aortic disorders (GA disorders) refer to a group of heritable conditions where the main artery of the heart, the aorta is affected. The commonality of GA disorders is the increased risk of serious cardiac complications such as an aortic tear or rupture. Affected individuals face multiple and complex medical and lifestyle challenges. Improving the individual’s understanding of the diagnosis, treatment, and associated impacts, is essential to facilitate coping, decision-making, and self-management. Evidence also show that patients who are more involved in the treatment of their chronic disease report improved health and psychological distress (Barlow et al., 2002). Currently available psycho-educational resources, however, are insufficient for those recently diagnosed with a GA disorder. The current thesis describes the development and evaluation of an evidence-based psycho-education booklet for patients recently diagnosed with a GA disorder.

Development: A patient psycho-educational booklet was developed based on a literature review, expert consultation, and guided by the National Health and Medical Research Council (NHMRC, 1999) recommendations.

Evaluation: A mixed method design was implemented. Twenty individuals with a diagnosis of a GA disorder, mean age = 49 years, equal number of males and females, with varying stages of disease and treatment, were recruited to complete the patient evaluation questionnaire. A further seven individuals were recruited for patient group
interviews. Nine clinicians with expertise in GA disorders completed the clinician evaluation questionnaire.

**Results:** The overall findings of the evaluation indicated that the information booklet was very well-received by the patient group and was considered informative, easy to read, practical, and a highly valuable resource. Overall, the clinicians endorsed the booklet as an extremely useful information resource about GA disorders and associated topics and supported giving the booklet to patients at the time of diagnosis and approved its ongoing use in consultations. The majority of clinicians commended the content as suitably pitched for the intended audience, and the format as appropriate in length, booklet size, layout and graphics.

**Conclusion:** This study aimed to develop and pilot evaluate a patient psycho-education booklet specifically for those recently diagnosed with a GA disorder. The evaluation showed that the booklet was considered very positively in its clarity, usefulness, and acceptability by the patients with a diagnosis of a GA disorder, and of usefulness, benefit, and acceptability by health professionals who provide medical support for patients with GA disorders. To our knowledge, this will be the first psycho-education booklet developed and evaluated for an Australian audience.
Chapter 1: Introduction

This chapter provides an overview of the conditions collectively known as Genetic Aortic Disorders (GA disorders), including their prevalence, aetiology, and treatment. The impact of GA disorders and the support needs of those with a chronic illness is also discussed.

1.1. Genetic Aortic Disorders

Genetic aortic disorders (GA disorders) are inherited conditions which affect the aorta, and aortic and mitral valves. The main conditions include Marfan syndrome (MFS), Loeys-Dietz syndrome (LDS), Thoracic Aortic Aneurysm and Dissection (TAAD), and Bicuspid Aortic Valve Aneurysm Syndrome (BAVAS). These conditions may lead to serious cardiac complications such as aneurysm and dissection, and mitral or aortic valve malfunction (Caglayan & Dundar, 2009).

GA disorders are known as autosomal dominant conditions. As seen in families affected by MFS, a child with one parent with the condition has 50% chance of inheriting the gene (Ammash et al., 2008). In other cases, however, genes are known to spontaneously mutate, known as a “de novo” mutation (Loeys et al., 2010; Pearson et al., 2008; Pomianowski & Elefteriades, 2013), with variations in the specific gene that is affected across the range of GA disorders.
Aortic Aneurysm and Dissection

The aorta extends from the heart through the chest and down into the abdominal area, dividing into blood vessels in each leg. There are four parts of the thoracic aorta: the aortic root; ascending aorta; aortic arch; and descending aorta. Due to structural weakness of the aorta, an aneurysm, a bulge or dilation in the wall of an artery can form (Hiratzka et al., 2010; Isselbacher, 2011). The pressure of blood inside the artery can then force the weak area to balloon outward resulting in the risk of rupture (Drinkwater, Kumar, & Taylor, 2010). Aortic aneurysms are classified depending on the shape, location, and formation. True aneurysms involve all three layers of the wall of the aorta (intima, media, and adventitia). Fusiform aneurysms appear generally as a symmetrical bulge, and saccular aneurysms are localised weakness of the artery wall. Thoracic aneurysms can occur along the aorta above the diaphragm, ascending aorta, aortic arch, and descending thoracic aorta, and those located under the diaphragm are classified as abdominal aneurysms (Drinkwater, Kumar, & Taylor, 2010; Hiratzka et al., 2010).

Aneurysms are generally diagnosed if the diameter is more than one and a half times the size of the normal aorta involving all layers of the arterial wall (Johnston, Rutherford, Tilson, Shah, Hollier, & Stanley, 1991). However, it is known that the aortic size varies due to various factors including age, gender, body size, method of measurement, type of imaging method used, and location of the measurement (Hiratzka et al., 2010). Using the aortic diameter indexed to height rather than aortic diameter alone to indicate surgical timing has been suggested for asymptomatic individuals with MFS or bicuspid aortic valve (Svensson & Khatin, 2002). Symptoms of an aortic aneurysm may include chest or
back pain (Isselbacher, 2005). The incidence rate of aortic dissection is estimated at 2.9 to 3.5 per 100,000 person (Ramanath, Oh, Sundt, & Eagle, 2009).

Aortic dissection and aneurysms may cause life-threatening situations where emergency medical intervention is required (Dietz & Pyeritz, 2002). The major cause of mortality and morbidity in GA disorders is due to aorta and valve complications (Ammash, Sundt, & Connolly, 2008; Loeys & Dietz, 2008; Pearson et al., 2008). However, age-adjusted mortality rates for all aortic aneurysms (including thoracic, abdominal, and thoracoabdominal) indicate that mortality has declined since 1990 to near four per 100,000 in 2006 (Centers for Disease Control and Prevention, National Center for Health Statistics, 2009).

Aortic dissection occurs when the inner layer of the aorta (intima) tear and separate from the middle layer (media), allowing blood to leak into the layers. As a result, this creates a second passageway for blood, known as the false lumen, in addition to the normal passageway, the true lumen. The false lumen may enlarge and subsequently block blood flow to or divert the blood flow away from the true lumen. The presence of an aortic aneurysm increases the risk of having an aortic dissection, but an aortic dissection can also occur in people with a normal sized aorta (Isselbacher, 2011; Drinkwater, Kumar, & Taylor, 2010; Hiratzka et al., 2010).

There are two main classification systems for aortic dissections based on the location of the tear in the inner lining. The DeBakey system classifies dissections into three types:
Type I – originating in the ascending aorta; Type II - originated in and is confined to ascending aorta; and Type III - originated in descending aorta. The Stanford classification system consists of two types: A - originating and involving the ascending aorta; and B - originating and involving the descending aorta (Hiratzka et al., 2010).

The treatment and management of GA disorders vary depending on the presenting features and may include a range of preventative, surveillance, and other strategies such as changes in lifestyle, cardiovascular monitoring, medical management, and pharmacological treatment (Caglayan & Dundar, 2009; Hiratzka et al., 2010). Modern aortic surgery includes a wide variety of techniques, including Dacron implants, stenting, and valve repairs and replacements (Hiratzka et al., 2010; Pearson et al., 2008). Guidelines for surgical intervention differ for the different GA disorder conditions and complications can occur below the recommended diameter, or may not occur at all for some individuals above the recommended dimensions (Ammash et al., 2008; Elefteriades & Farkas, 2010). Medications such as beta blockers have been used to treat aortic dissections to lower blood pressure and reduce the heart rate. In MFS, beta blockers have been shown to slow aortic root growth and prevent the aortic dissection from worsening which can lengthen the time to surgery (Ammash et al., 2008). Shores, Berger, Murphy, & Pyeritz (1994) reported that beta blocker therapy (propranolol) in comparison to no use of beta blocker resulted in slower rate of aortic dilatation, and lower mortality. However, these results cannot be generalised across to other GA disorders at this stage (Isselbacher, 2005).
Lifestyle recommendations include avoidance of competitive or high impact contact sports, which can increase the risk of acute aortic dissection, although moderate exercise and other activities are recommended based on the individual’s risk profile (Hiratzka et al., 2010; Raanani & Ghosh, 2008). Substances that place increased stress on the aorta, including cigarette smoking, cocaine and methamphetamines use, should be avoided (Booher & Eagle, 2011).

Due to the advances in diagnosis and management of GA disorders, treatment outcomes and life expectancy have improved (Pearson et al., 2008). Despite this progress, GA disorders present the need for complex treatment decisions and lifelong planning and intervention.

1.1.1. Marfan Syndrome (MFS)

MFS occurs as a result of mutation of the fibrillin-1 gene (FBN1) on chromosome 15q21 (Dietz et al., 1995). Defects of fibrillin results in connective disorders affecting the aorta, lens of the eye, cartilage, ligaments, and the pulmonary pleura (Judge & Dietz, 2005; Loeys et al., 2010). Features of MFS include aortic dissection, aortic aneurysms, detached retina, pneumothoraces, arachnodactyly, scoliosis, and sternal deformities (William, Davies, Stuary, & Fraser, 2008). The manifestation of MFS differs widely across affected individuals from mild to very severe problems. Some individuals may present with aortic problems, while others may have ophthalmic or orthopaedic issues. Individuals with MFS often have physical features such as long thin limbs, highly arched palate and crowded teeth, small lower jaw, scoliosis, and a thin, narrow face (Doyle &
The Development and Evaluation of a Psycho-education Booklet for Genetic Aortic Disorders

Dietz, 2011). The signs of MFS can be present from a young age, with aortic dissection or aortic aneurysms occurring in young adulthood (Judge & Dietz, 2005).

Diagnosis of MFS is based on the revised Ghent Nosology criteria (Loeys et al., 2010). The criteria places emphasis on cardiovascular indicators, with aortic root aneurysm and ectopia lentis as the principal clinical features. Magnetic resonance angiogram (MRA) or computerised tomogram (CT) scans are used to detect abnormalities of the aorta from the aortic valve to the bilateral common iliac arteries. Other assessments include orthopaedic and ophthalmologic evaluations. Information about a family history of MFS and other connective tissue disorders is important due to the heritability issue (Peters, Kong, Horne, Francomano, & Biesecker, 2001) and genetic counselling can be offered.

The prevalence of MFS is estimated at 1 per 5000 individuals, with no gender differences (Ho, Tran, & Bektas, 2005; Judge & Dietz, 2005). About 75% of affected individuals inherited the condition, and 25% are thought to be due to spontaneous genetic mutation (Loeys et al., 2010; Pearson, et al., 2008; Summers et al., 2006). MFS affects males and females equally, with no specific patterns across ethnicity.

Treatment of MFS ideally involves a multidisciplinary approach (Ammash et al., 2008), including active surveillance of the aorta and aortic valves, antihypertensive therapy, avoidance of high impact activities, and management of other associated issues. Planned surgery may be considered depending on the risk associated with the aortic aneurysm. Pregnant women with MFS require regular monitoring given the increased risk of aortic
dissection during delivery and three month post-partum (Goland & Elkayam, 2009). With beta-blocker therapy and elective surgical repair, the median probability of survival is over 72 years for individuals with MFS (Silverman & Burton, 1995).

1.1.2. Loey-Dietz Syndrome (LDS)

Loey-Dietz Syndrome (LDS) manifests with similar physical features to MFS and was first described in 2005 (Aalberts et al., 2008; Loeys et al., 2005). LDS is caused by mutations in the TGFBR1 or TGFBR2 genes that affect the signals in the cells that promote tissue growth and development. The alteration results in the disorganisation of the elastic fibres in the aortic wall, and is associated with a greater likelihood of tears and dissections than MFS (Summers et al., 2006; Van Hemelrijk et al., 2010). Consequently, treatment recommendations include annual MR imaging and surgical management of the aneurysm can be warranted (Hiratzka et al., 2010).

The main characteristics of LDS include twisting or spiralled arteries in neck vessels, widely spaced eyes, and broad or split uvula. Other features such as cleft palate, velvety and translucent skin, and clubbed feet are features in LDS but not in MFS. It is estimated that 25% of cases of LDS are inherited, with around 75% occurring as a result of a spontaneous genetic mutation. LDS is estimated to affect a smaller number of individuals than MFS (Aalberts, et al., 2008).
1.1.3. Thoracic Aortic Aneurysm and Dissection (TAAD)

A thoracic aortic aneurysm can lead to a tear in the artery wall that causes bleeding or dissection. TAAD is commonly asymptomatic, and is frequently discovered when dissection or rupture of the aortic aneurysm occurs. Therefore, many individuals with TAAD are diagnosed as a result of emergency medical intervention (Coady et al., 1999; Loeys & Dietz, 2008).

Treatment of detected TAAs involve life-long monitoring, and may include planned or emergency surgical treatment. Slow growing and small thoracic aortic aneurysms are at less risk of rupture in comparison to fast growing TAAs. There are no differences in the incidence between genders, but TAAD increases in frequency with older age (Clouse, Hallett, Schaff, Gayari, & Ilstrup, 1998). The incidence rate of thoracic aortic aneurysms is estimated at approximately 10.4 per 100,000 person-years (Ramanath, Oh, Sundt, & Eagle, 2009).

1.1.4. Bicuspid Aortic Valve Aneurysm Syndrome (BAVAS)

A bicuspid aortic valve, which lies between the heart and the aorta, has two instead of the usual three leaflets that serve to regulate blood flow. The underlying connective tissue disorder also weakens the aortic tissues, and can be associated with weakening of the aortic wall, leading to an aneurysm forming. Approximately one to two in 100 people have a bicuspid aortic valve, of whom half will have some aortic dilation (enlargement) (Guo et al., 2007). Approximately 9% of patients with bicuspid aortic valves have other affected family members (Huntington, Hunter, & Chan, 1997).
Summary

The conditions collectively known as GA disorders affect the aorta, and aortic and mitral valves, although features manifest differently in each individual. Treatment may consist of planned or unplanned surgery, medication management and lifestyle changes. The impact of a GA disorder may affect a range of areas of an individual’s life, and accepting the restrictions that the condition may require can be particularly difficult if considerable lifestyle changes are needed (Livneh & Antonak, 2005). In Velvin and colleagues’s (2015) systematic review, it was highlighted that individuals with MFS may benefit from psychosocial support to assist them to cope with the medical and psychosocial impact.

1.2. Psychosocial Impact of GA Disorders

The psychosocial impact of a GA disorder or other chronic illness refers to the psychological, social, and physical functioning, and quality of life of the individual. Individuals affected by GA disorders face multiple and complex challenges across various areas of their lives. The affected individual needs to make permanent adjustments to their lifestyle and self-care (e.g. avoiding contact sports, life-long medication use), make difficult decisions regarding future plans (e.g. reproductive decisions, screening of family members/children), and cope with the ongoing psychosocial impact of the condition (e.g. uncertainty, psychological impact such as stress, anxiety, and depression, and stigma). It has been shown that chronic illness can be disruptive to work, daily life activities, and social roles, and the experience of treatments and progression of the disease can be traumatising (Livneh & Antonak, 2005). In addition, Rand-Hendriksen and colleagues (2007) reported reduced scores for health-related quality of life in
individuals with MFS to be comparable to individuals with scores in other chronic conditions and disabilities such as uveitis, hypertrophic cardiomyopathy without angina, cystic fibrosis, and Behçet Syndrome.

The available literature on the psychosocial impact of GA disorders is mainly centred on individuals with MFS, presumably because it is the more recognised, common, and visible GA disorder. Nonetheless, research on the psychosocial impact of MFS is highly relevant across the spectrum of GA disorders given the commonalities in the features, treatment, and potential consequences of the conditions.

1.2.1. Illness Uncertainty

A diagnosis of a GA disorder is likely to raise many and various concerns for the affected person and their family/support persons. Uncertainty relating to the diagnosis, treatment, and the impact on their daily life and future can be a significant psychosocial stressor. Illness uncertainty in chronic illnesses refers to the distress experienced as a sense of loss of control and perceived varying state of doubt (Wright, Afari, & Zautra, 2009).

Uncertainty in chronic illness, and the resulting difficulty in adjustment, can occur when there is uncertainty in the areas of diagnosis or prognosis, when an illness is either sudden and novel, or chronic where there is unpredictability in the disease course (Mishel, 1999). The uncertainty can be influenced by insufficient information about the diagnosis or severity of the illness, and difficulties relating to the health care system (Mishel, 1999). In particular, with GA disorders, complications that may arise can be potentially life-threatening, and unexpected surgery or other emergency intervention means that the
patient is unable to anticipate and prepare for the short and long term outcomes such as the potential loss of physical functioning and lifestyle restrictions.

Chronic illness also commonly creates uncertainty due to its impact on the body, sense of self, and social aspects of life (Faircloth et al., 2004). Some research has suggested that those with a recent diagnosis of a chronic illness experience higher levels of illness uncertainty than those further along in time since diagnosis due to the increased knowledge and illness management skills over time, however, some individuals do continue to experience great uncertainty (Mishel, 1990; Bailey & Neilsen, 1993). The theory of illness uncertainty proposed by Mishel (1990; 1999) explains that adjustment to acute and chronic illness depends on the individual’s appraisal of the uncertainty. If uncertainty is perceived as an opportunity for a more desirable outcome, then an individual is more easily able to achieve positive adaptation to the illness, whereas if uncertainty is perceived as a threat, then maladaptive coping, higher psychological distress, and reduced quality of life can occur (Mishel, 1999; Wright, Afari, & Zautra, 2009). In a study by Connors, Jeremy, Fisher, Sharpe, & Juraskova (2015) that explored coping and adjustment processes of individuals with a GA disorder, acceptance of uncertainty due to the condition helped individuals to move their focus from aspects that had low control to aspects that could be controlled. The literature suggests that individuals diagnosed with a GA disorder may experience initial and ongoing uncertainty from the time of diagnosis, particularly with complications and associated interventions that may occur, and must manage the uncertainty that these events bring, along with other
psychosocial impacts including psychological, social, physical restrictions, vocational, and quality of life.

1.2.2. Psychological Impact

It has been shown that individuals with chronic illnesses may experience psychological distress relating to loss and grief, stigma, shock, anxiety, and depression (Livneh & Antonak, 2005). In the MFS literature, Velvin, Bathen, Rand-Hendriksen, and Geirdal (2015) found in their systematic review that there were greatly varying levels of psychological distress reported amongst individuals with MFS in comparison to the general population. Peters, Horne, Kong, Francomano, & Biesecker (2001) reported that 44% of their sample of 174 individuals with MFS experienced clinical levels of depression as measured by the Center for Epidemiologic Studies Depression Scale (CES-D). Whereas Van Togerloo & De Paepe (1998) reported no significant levels of depression or anxiety with their sample of seventeen adolescents and young adults who completed the State-Trait Anxiety Inventory. Divergent results from studies on psychological distress may result from small sample sizes, a variety of methodologies, and the use of different psychological scales.

It has however, been reported that emotional distress may also be related to issues such as stigma, physical limitations, the perception of difference to others, and experiences of life-threatening complications (Rand-Hendriksen, Johansen, Semb, Geiran, Stanghelle, & Finset, 2010; Peters, Kong, Hanslo, Francomano, Biesecker, 2002; Schneider, Davis, Boxer, Fisher, & Friedman, 1990; Van Togerloo & De Paepe, 1998). Further, it has been
identified that individuals with MFS cope better with their diagnosis when they had accurate knowledge of their diagnosis and a realistic expectations of outcomes (Schneider, Davis, Boxer, Fisher, & Friedman, 1990; Giarelli, Bernhardt, Mack, & Pyeritz, 2008). It is evident that the impact of GA disorders on psychological distress and coping is influenced by a range of factors. It is important that individuals affect by a GA disorder be aware of and have information about the possible psychological impact, and that individuals are supported to access intervention should it be required.

1.2.3. Social Impact

Social stigma experienced by individuals with MFS has been reported in various studies. In a study of 174 adults with MFS conducted by Peters, Apse, Blackford, McHugh, Michalic, & Biesecker (2005), 32% of respondents of a survey reported feeling discriminated against or socially devalued due to having MFS, leading to social withdrawal from social situations where stigmatisation was anticipated. Educating others about MFS was suggested by over half of the respondents as a means of addressing stigma. Van Tongerloo and De Paepe (1998) utilised semi-structured interviews and standardised psychological questionnaires in a study of adolescents and young adults with MFS. Respondents reported that they were teased by peers during their childhood due to the physical features and limitations common in MFS, and were more likely to consider themselves as introverts. Respondents also reinforced the need for accurate medical information about MFS and psychological support. These needs are likely highly relevant and important across all GA disorders.
The impact on other social aspects described in the literature include intimate relationships and family life. De Bie et al. (2004) reported that having MFS had a negative influence on the individual’s sexual relationship due to the perception of their own body image. Having a diagnosis of MFS was also reported to have an influence on the decision to have children. Concerns about reproduction included the risk of passing on the condition to their children, and risk to their own health such as aortic rupture or dissection (Peters et al., 2001; De Bie et al., 2004; Van Tongerloo & De Paepe, 1998). However, De Bie et al. (2004) and Peters et al. (2001) also reported that the many individuals with MFS considered themselves living normal family lives and had children. Given that there is the potential impact on reproductive decisions for those affected by a GA disorder, it is essential that there is accurate information about the risks and options. Information or educational resources relating to GA disorders should provide evidence-based medical information and acknowledge the potential social impact including genetic concerns, and to guide patients on how/where to find support services.

1.2.4. Vocational Impact

Physical restrictions associated with a GA disorder may limit an individual’s vocational functioning, therefore impacting on vocational choices or opportunities. In a systematic review of the psychosocial aspects of MFS conducted by Velvin, Bathen, Rand-Hendriksen, and Geirdal (2015), it was found that individuals with MFS reported that physical limitations impacted on their capacity to carry out daily functions, restricted their work opportunities and negatively impacted on their social interactions. For school-aged individuals, their school attendance was reduced.
The limitations that have been reported to negatively impact on career or work choices include the need to restrict physical exertion and reduced working capacity due to the GA disorder, therefore limiting work choices (Fusa-Poli et al., 2008; De Bie et al., 2004). Peters et al. (2005) showed that individuals with MFS reported remaining in a workplace despite job dissatisfaction, had experienced perceived discrimination, and felt they had limited occupational options. Further, De Bie et al. (2014) found that the perception of reduced capacity for work due to MFS increased with older age, and occurred at an earlier age than for the general population (40% for those over 40 years). There have been no findings that indicate a relationship between work/vocational difficulties and the severity of the GA disorder. Considering the potential impact of a GA disorder on vocational functioning, it is essential that patients have information about possible limitations and options and to be supported in discussing their concerns with their clinician. It is important that a psycho-education resource include information about possible vocational impacts and support to manage these issues.

1.2.5. Health-Related Quality of Life

There is limited and conflicting information about the overall impact of MFS and other GA Disorders on quality of life (QOL). Rand-Hendriksen, Johansen, Semb, Geiran, Stanghellie, & Finset (2010) reported reduced health-related QOL scores overall in 84 adults comparison to the general population in Norway as measured by the Short Form 36. Verbraecken, Declerck, Van de Heyning, De Backer, & Wouters (2001) also found lower QOL scores in 15 adults with MFS on the Health-Related Quality Of Life
questionnaire in comparison to age matched controls, except in psychological and social functioning, and energy levels. However, Peters et al. (2002) reported no significant difference in QOL in 174 adults with MFS in comparison individuals with other cardiovascular disease as assessed by the Ferrans and Powers Quality of Life Index, Cardiac Version III, although the QOL scores were significantly lower in the psychological subscale. A study by Fusar-Poli et al. (2008) with a sample of 36 adults with MFS reported similar findings using the Short Form (36) Health Survey (SF-36). The contrast in findings as described above may be a result of relatively small sample sizes in the studies, differing measurement tools used, and divergent characteristics of the sample populations. There is a need for further research on the impact of GA disorders on health-related QOL, and subsequently how these issues can be addressed through various means including through patient health education strategies.

1.3. Adherence to treatment in Chronic Illness

The term medical adherence refers to the congruence between the patient’s behaviour and the clinician’s recommendations that were agreed upon (Brunton, 2011). Non-adherence to medical advice is a major area of concern in chronic illnesses as poor adherence contributes to disease complications, disruptions in lifestyle, and increased hospitalisation (Berg, Evangelista, & Dunbar-Jacob, 2002). It has been shown that non-adherence affects disease progression and increases morbidity and mortality (McHorney, 2009). The estimated rate of adherence to medical recommendations in chronic illnesses has been found to be approximately 50% (Dunbar-Jacob et al., 2000). Non-adherence to treatment also appears to be a major concern in individuals with a GA disorder.
Various MFS studies have identified that a proportion of patients do not follow medical recommendations. In relation to lifestyle changes with GA disorders, restrictions on high-impact activities is recommended. However, it has been found that a small proportion of individuals with MFS continue to engage in high-risk activities (De Bie, De Paepe, Delvaux, Davies, & Hennekam, 2004). Van Tongerloo & De Paepe’s study (1998) found that several participants reported accepting the diagnosis but continued playing high impact sports against medical advice. Peters et al. (2001) reported that those who followed exercise recommendations were more likely to have a greater negative view about the consequences of MFS. In relation to why some individuals may not follow recommendations, De Bie et al. (2004) suggested that the potential consequences of asymptomatic cardiac complications may be overlooked or more easily discounted despite patients having knowledge of the risks.

In GA disorders, the use of medications as prescribed is essential for maintaining well-being. In a study of 22 young persons (age range 11-24 years) with MFS, it was found that their medical compliance was at suboptimal levels (Schneider, Davis, Boxer, Fisher, & Friedman, 1990). These compliance levels were similar to teenagers with other chronic illnesses. Reasons for non-adherence to medication recommendations included parental disapproval of medication use and avoidance of having the prescription filled. However, in another study, adherence to medication recommendations was found to be at 80% with adults with MFS, and physical activity was chosen in consideration of their diagnosis and recommendations (Peters et al., 2001). The researchers found that individuals who believed that medications may have harmful effects and significant consequences with
inappropriate use were more likely to adhere to the recommended medication usage. These studies suggest that medication adherence is influenced by beliefs about the effects (both positive and negative) of medication and support or lack of support from significant others for its use.

Adherence and self-management of chronic illness including GA disorders requires the development and implementation of skills including knowledge seeking, implementing healthy action, and life-long self-monitoring (Giarelli, Bernhardt, Mack, & Pyeritz, 2008). Research shows that relevant and quality information helps with patient’s understanding, coping, and decision-making in individuals affected by cancer (Iconomou, Viha, Koutras, Vagenakis, & Kalofonos, 2002). A variety of strategies to promote adherence and self-management have been recommended, including improving patient-clinician communication, patient education, and delivery methods of information (Brunton, 2011).

1.4. Coping and Adjustment to a Diagnosis of GA Disorder

There is very limited literature on coping and adjustment in individuals diagnosed with a GA disorder. However, a recent qualitative study by Connors, Jeremy, Fisher, Sharpe, & Juraskova (2015) presented important information on the psychosocial impact, coping, adjustment, and unmet informational needs in individuals with these conditions. The study identified factors that influenced treatment adherence and psychological outcomes,
using a battery of questionnaires and semi-structured telephone interviews to elicit information on how individuals coped and adjusted to a diagnosis of GA disorder. The information examined included information and decision-making preferences, levels of psychological distress, and coping styles.

This study consisted of a sample size of twenty-one adults (mean age 38.8 years) with the time since diagnosis ranging from one to greater than eleven years. Results from the interviews identified five major themes relating to how the respondents adjusted and coped from the time of diagnosis of a GA disorder. A model of adjustment was developed that illustrates the influences of these experiences on psychosocial outcomes and treatment adherence.

The central concept identified in Connor’s et al. (2015) study was the process of adjustment identified as Negotiating the narrative of self and the GA disorder which encompassed five main themes and associated subthemes. The main theme of “restrictions upon lifestyle” related most closely to adherence as it described that individuals accepted treatment and limitations of having a GA disorder as “part of life”, whereas low acceptance was observed in individuals who “wanted to be normal”. “Negotiated the restrictions” occurred when the individual sought advice on and developed an understanding and expectations of their physical limitations.

The second theme of “loss of control” related to the emotional challenges following surgery, with “fear of the unknown” describing distress occurring prior to surgery about
the possible outcomes. The “impact of the GA disorder” included the outcomes and side-effects of surgery and medication that may have contributed to a sense of reduced bodily control as well as physical restrictions during recovery from surgery. “Fear of death” for a small proportion of the respondents was similar to a traumatic response to a life-threatening situation. “Gaining control” described the process of regaining stabilisation that involved practical strategies, cognitive shifts, and an optimistic personality trait.

The third theme of “future” described both uncertainty about the future, and transformation and hope. The grief that occurred with an imagined future and subsequent change in perception of self was described as “loss of imagined future”. “An uncertain future” referred to the unknown outcomes, and “transformation and hope” described positive changes in perspectives and identity as a result of the treatment process.

The fourth theme of “support” included: “health professional support”; “emotional and practical support”; “spiritual support”; and “support for carers”, highlighting the importance of trust in and continuity of care from the primary medical clinicians, the impact of psychosocial support on wellbeing, and the recognition of unmet emotional and physical needs of carers.

The fifth theme of “unmet needs” is of particular relevance to the current study as described in this thesis. The subtheme of unmet “information needs” demonstrated the need for accurate, easily accessible, specialist clinician endorsed information about GA disorders, treatment, psychosocial impacts, and further resources. The subtheme of
“connect with peers” shows that individuals with a GA disorder want to be able to share others’ experiences and their own in order to normalise their experiences and to assist others through their journey.

The results of the Connors et al. (2015) study showed that individuals with a GA disorder may experience a range of psychosocial impacts from the time of diagnosis. An integrated model of adjustment to GA disorders was developed by the researchers which illustrates how the identified themes and processes can influence adherence and psychological outcomes. This includes the contextual factors influencing the appraisal of the GA disorder threat, how the individual appraises the threat, and then the different pathways to adherence and outcomes. The model is provided in Figure 1.1. Generalisability of the model however, may be limited due to the study’s small sample size and possible recruitment bias that may have resulted from the use of a single site. However, the study produced extremely useful insights and valuable information about the unmet needs, psychosocial impact, coping and adjustment processes with individuals with GA disorders.
Figure 1.1. Integrated model of adjustment in GA disorders (Connors et al., 2015)

GA THREAT
(abstract or concrete)

CONTEXTUAL FACTORS
- Narrative of self
- Narrative of illness
- Supports
- Stressors

APPROVAL OF THREAT
- Can narrative of self and GA disorder be positively integrated?

Is appraisal of vulnerability to threat realistic?

Yes

Try to change illness narrative
- Downward comparison
- Reframing
- Accept uncertainty
- Information seeking
- Control controllable

Integrated narrative of self

Try to change narrative of self
- Response shift
- Reimagining future
- Transformation
- Imbuend meaning

Integrated narrative of self

No

Try to keep narrative of self separate from GA disorder
- Disengagement strategies

Try to keep narrative of self separate from GA disorder

Combined

Optimistic traits

Good adherence
Low psychological distress

Poor adherence
High psychological distress

Good adherence
High psychological distress

Poor adherence
Low psychological distress

Poor adherence
High psychological distress
The results of the Connors et al. study showed a distinct gap in information resources for individuals with a GA disorder. Particular suggestions from the participants about what they would want in information resources included information specific to the condition and lifestyle impact, and that individuals wanted resources developed by health professionals to aid in the understanding of the condition and treatment. A suggested mode of information delivery was for printed information, containing pictures of the heart and aorta, with medical terms and signs and symptoms explained, an emergency plan, and suggestions on where to get more information.

Several other studies have also demonstrated unmet information needs of individuals with a GA disorder. From their study of individuals with MFS aged between 16-35 years of age, Van Tongerloo and De Paepe (1998) reported that most highlighted that accurate information about the condition immediately post-diagnosis was important, and that psychological support was helpful. Similarly, Wachters-Kaufmann, Schuling, and Meyboom-de Jong (2005) reported that after a stroke, patients and caregivers preferred to receive information within 24 hours, and preferred information in written form.

**Summary**

The conditions collectively known as GA disorders affect the aorta, and aortic and mitral valves, putting the affected individual at risk of an aortic tear or rupture. Individuals with a GA disorder face multiple and complex medical and lifestyle challenges as well as a range of psychosocial impacts. Improving the individual’s understanding of the
diagnosis, treatment, and associated impacts, is essential to facilitate coping and
adjustment, decision-making, treatment adherence and self-management. In light of the
review of the psychosocial impact and coping processes as presented above, it is evident
that effective methods of information dissemination, and improving the accessibility of
information is required to meet the needs of individuals affected by GA disorders. An
information resource such as a psycho-education booklet specifically about GA disorders
would be an important approach in addressing unmet information needs in this
population.

The next chapter will present a review of patient information needs following diagnosis
of a chronic illness, the purpose and objectives of health information, developing health
information resources, and evaluation of resources.
Chapter 2: Patient Information Needs

In this chapter, a review of the information needs of persons with chronic conditions will be presented. Due to the very limited literature on patient information needs on GA disorders, the review will encompass a range of chronic conditions given their similarities in that: GA disorders and other more common chronic conditions are prolonged in duration; complex and varied in their nature; do not spontaneously resolve; and are rarely completely cured. The chronic conditions of interest include cardiovascular diseases, cancers, chronic respiratory diseases, diabetes, and different forms of arthritis. The purpose and objectives of written health information resources, and the development and evaluation processes of resources will also be outlined. This chapter will conclude with the aims of the current study.

2.1 Information Needs of Individuals with Chronic Conditions

2.1.1 Patient Health Education

Patient education is an important means of addressing the increasing burden of chronic disease, in order to encourage more active self-management of medical problems (Holman & Lorig, 2000). It has been postulated that a major barrier to adequate self-care in people affected by chronic illness is the lack of relevant health information (Blickem et al., 2011).

Research has shown that patients often do not have a good understanding of their diagnosis, prognosis, treatment intent and options (Hagerty, Butow, Ellis, Dimitry, &
Tattersall, 2005). In a study examining 62 adult patients’ knowledge of congenital heart disease, a poor level of understanding (less than 50% correct responses) about the reasons for follow up, symptoms relating to deteriorating heart disease, and heritability of the condition was reported (Moons et al., 2001). Similarly, in a cross-sectional survey conducted by Lesch, Specht, Lux, Frey, Utens, and Bauer (2014) involving 596 participants with congenital heart defects across three main age ranges (10-30 years old), a major knowledge gap in relation to the condition and self-management was shown.

It has been reported however, that a better understanding of treatment options can influence treatment choices and promote adherence (West & Baile, 2010), and that patients who are better informed about their illness experience fewer medical problems (Hermiz et al., 2002; Ryan et al., 2003). Evidence shows that patients who are more involved in the treatment of their chronic disease and participate in shared decision-making report better coping, increased self-care, better treatment adherence, and less psychological distress (Barlow et al., 2002; Perreda et al., 2008). Shared decision-making is defined as the communication between clinician and patient in the context of decision-making that is supported by best available evidence that informs patients of screening, treatment or management options, likely risks and benefits, taking into consideration the patient’s values and preferences (Stiggelbout, Van der Weijden, De Wit, Frosch, Légaré, Montori Elwyn, et al., 2012; Elwyn, Laitner, Coulter, Walker, Watson, & Thomson, 2010). It is known that shared decision-making is facilitated by the provision of health information and good patient-clinician communication within the chronic illness setting (Kanj, & Mitic, 2009). With GA disorders, shared decision-making is important given
that the affected individual will require life-long medical care to prevent complications
and to maintain health. A psycho-education booklet on GA disorders can facilitate an
individual recently diagnosed to make sense of often complex medical information and
treatment recommendations, and support the individual to communicate their concerns or
questions to their clinician.

2.1.2 Health literacy and Patient Knowledge of their Condition

Health literacy is a term that refers to a combination of skills that enables individuals to
understand and utilise information to achieve and maintain good health. There are various
definitions of health literacy that are in use in the current literature. One of the broadest
definitions of health literacy is from the World Health Organisation: “The cognitive and
social skills which determine the motivation and ability of individuals to gain access to,
and understand and use information in ways which promote and maintain good health”
(Smith, Tang, & Nutbeam, 2006). Defined more specifically, Kickbusch, Wait, and Maag
(2012) proposed that health literacy encompasses individuals’ ability to make health
decisions in a variety of contexts including “at home, in the community, at the workplace,
in the healthcare system, the marketplace and the political arena” (p.8). According to
Nutbeam (2000), the role of health information and education in improving health
literacy involves three main goals and their associated activities: i) improvement of basic
or functional health literacy through provision of patient information leaflets; ii)
promotion of interactive or participative skills through skills development programs; and
iii) facilitating critical or analytic skills through community-based development programs
that promote change at a policy and/or organisational level. The development of a
psycho-education booklet as described in this thesis supports the first goal of promoting health knowledge, communication with health professionals, and facilitating decision-making and self-management.

In the context of GA disorders, Coulter & Ellins’ (2006) definition would seem most appropriate – namely, that health literacy is the capacity to apply a range of functional skills, including general literacy skills, in the context of healthcare, in order for individuals to be involved in and make decisions about their treatment. It has been shown that good health literacy facilitates self-management in chronic illnesses, as it involves the understanding often complex medical information, development of treatment plans, lifestyle adjustments, and navigating the health care system (Lloyd, Ammary, Epstein, Johnson, & Rhee, 2006).

The Australian Bureau of Statistics (2009) reported on health literacy in 15-74 year olds in Australia, measured by the Adult Literacy and Life Skills Survey (ALLS). It was found although 41% of those assessed had adequate or better health literacy skills (e.g. able to combine information from text and graph formats to correctly assess the safety of a health product), 40% of the general population had skills below the minimum level considered necessary for meeting the complex demands of everyday life (e.g. difficulty with locating information on a bottle of medicine about the maximum number of days the medicine could be taken). It is therefore expected that a proportion of the patient population with chronic illnesses, including GAD patients, would have considerable difficulty with health literacy.
The literature shows that patients with chronic illnesses have significant gaps in knowledge of their condition due to poor health literacy. In a study examining the relationship between health literacy and knowledge of disease in patients with a chronic illness, it was found that almost a quarter of patients had lower levels of health knowledge due to difficulties with literacy than those with adequate health literacy skills (Gazmararian, Williams, Peel, & Baker, 2003). In the rheumatoid arthritis literature, of 80 individuals with rheumatoid arthritis who were assessed for level of functional health literacy, 10% were found to have difficulty with reading and interpreting health texts such as simple written instructions or prescription labels (Buchbinder, Hall, & Youd, 2006). It is of concern that a proportion of the general population have difficulties with the comprehension of health information which may lead to lower rates of health screening or diagnostic testing, and poorer involvement in the health care system (Australian Bureau of Statistics, 2009). With diagnostic screening and other ongoing investigations being of particular importance in GA disorders, it is important to ensure that the information in a psycho-education booklet on GA disorders is tailored to the broadest level of health literacy.

In sum, it is evident that individuals need to have adequate skills to be able to read, understand, and use healthcare information in effective ways. Better health literacy will help individuals to determine what good health information is and then apply the information in their own situation (Jordan, Briggs, Brand, Osborne, 2008). A patient-centred information resource, such as a psycho-education booklet could facilitate the acquisition and retention of knowledge, particularly for uncommon conditions such as
GA disorders. Important elements that would facilitate better understanding of the information would include the appropriate use of lay language for the target audience, using a variety of tools such as diagrams and graphics to support the text, and important points highlighted.

**Readability**

For text-based information to be effectively communicated, the information needs to take into consideration its readability and comprehensibility based on the demands on literacy skills. The readability of text can be assessed through various formulas, including the Simple Measure of Gobbledygook (SMOG) formula (McLaughlin, 1969), the Flesch Reading Ease (Flesch, 1948), and Fry formula (Fry, 1968). In general, these formulas assign an equivalent reading grade level and a score for reading difficulty based on the number of difficult words (syllables in a word), and the number and average length of sentences.

It is reported that 40% of the Australian population have some, or considerable, difficulty with written health information material (Australian Bureau of Statistics, 2009). Yet, reviews of patient health resources show that they are written at a reading level of eighth-to twelfth-grade level (US) which is above the recommendation for the reading level at or below the sixth- to eighth-grade level (Nielsen-Bohlman, Panzer, & Kindig, 2004). Smith, Gooding, Brown, and Frew (1998) estimated that 97% of asthma pamphlets used in general medical practice would not be comprehensible to the majority of the British population. Cervical cancer prevention materials were also found to have a readability level too high for the majority of users (Helitzer, Hoolis, Cotner, Oestreicher, 2009).
Such materials demand high literacy skills, and often have ambiguous, technical, or dense text.

It has been suggested however, that using readability formulas does not adequately evaluate the comprehensibility of written materials as other factors, such as tone, framing, timing, terminology, and interest and motivation of the reader, which also influences the usefulness of the resources (Eysenbach et al., 2002). Caution must also be taken with the use and interpretation of readability formulas as they do not take into consideration the reader’s familiarity with medical terminology, particularly as patients acquire knowledge and increase their medical vocabulary over time, as is the case with chronic illnesses (Smith, Gooding, Brown, & Frew, 1998; Moult, Franck, & Brady, 2004). In these cases, the readability score may underestimate the readability of the text. Therefore, basing the text on a readability score alone without considering the factors will not necessarily improve the comprehensibility and usefulness of the material. Other influencing factors such as language, tone, framing, content, format and design of written resources will be described in the next chapter with the description of the development of the study booklet.

2.1.3 What patients want to know about their medical condition

Studies have shown that the most common information needs of patients include information about an illness or medical condition, diagnosis, and treatment options (Clarke et al., 2015; Coulter & Ellins, 2006). In a multi-site survey of 2331 cancer patients, 87% of participants indicated that they wanted both good and bad news (Jenkins,
The Development and Evaluation of a Psycho-education Booklet for Genetic Aortic Disorders

Fallowfield, & Saul, 2001). Ivarsson, Larsson, Lührs, and Sjöberg (2005) reported that patients surveyed prior to cardiac surgery (n=182) who had received detailed written information about possible complications were significantly more satisfied with the information both pre- and post-operatively, compared to those who received standard care.

It is also known that patients want more information than they commonly receive (Coulter & Magee, 2003). The literature on information needs of individuals with cardiac-related conditions and/or whom have experienced surgery, consistently shows that patients want more specific information about their condition, treatment options, self-management recommendations, and how to manage psycho-social issues. In a qualitative study conducted by Pier and colleagues (2008), fourteen individuals who had experienced myocardial infarction, coronary artery bypass graft surgery, angioplasty or angina, rated that their most important information needs were the identification and management of risk-related physical symptoms, and psycho-social information relating to enhancing their social support. Similarly, in a cross-sectional survey of 596 participants with congenital heart defects aged 10-30 years, it was found that across the age ranges, unmet information needs included psycho-social and lifestyle issues such as potential impact of their condition on school/work, career and sports (Lesch et al., 2014).

Individuals at different stages of illness or life stages have different priorities and therefore information needs. In studies investigating demographic differences and information preferences, it has been found that higher educational level and younger age
are positively associated with higher health information seeking (Rutten et al., 2005). Age may also influence the type of information sought, with younger patients wanting more information about sexuality and physical appearance (Davison et al., 2002; Degner et al., 1997; Luker et al., 1995), and also younger patients preferring more treatment-related information than older patients (Jenkins et al., 2001).

Another issue relating to unmet information needs is that there can be discrepancies between health professionals' and patients' views on information needs (Capirci et al., Adab and colleagues (2004) reported that patients with arthritis (n=201) viewed information about medication side effects, diet and alternative treatments as a greater unmet information need in comparison to health professionals (n=232). Further, a study by van Weert, van Dulmen, BaÈr, and Venus (2003) found insufficient content and level of information provided in consultations between cardiac surgery patients and health professionals. Preoperative consultations between fifty-one cardiac surgery patients and a range of health care providers (including physicians, nurses and health educators) were video-taped. The verbal interaction between each patient and health care provider was analysed using a coding system (Roter Interaction Analysis System) consisting of thirty-two categories. A checklist pre-determined by the research team, including one hundred and twenty-three specific categories of information, was also used to analyse the content of the patient-health professional communication. Findings show that there was little provision of psycho-educational information and emotional support by health professionals. The psycho-education categories included information about the possible psycho-social consequences of cardiac surgery, and checking by the health professional
about the patient’s understanding of information. Emotional support categories included whether the health professional asked about the patient’s feelings, worries or concerns about surgery, offered verbal demonstrations of empathy, or gave reassurance. Analysis of the communication from patients showed that they mostly sought information about short-term, practical information, and minimally about psycho-social impact. The researchers suggested that a greater focus on psycho-social issues by health care providers may facilitate patients to express their emotional and other psycho-social needs. The gap between what information patients want and what clinicians believe is necessary or has been provided, can lead to misunderstandings about treatment and subsequently poor or non-adherence to treatment plans (Britten et al., 2000). This information is of particular importance in the GA disorders population given that patients may already have undergone surgery, or may face the ongoing possibility of surgery.

In sum, the literature demonstrates that there appears to be major gaps in knowledge in persons affected by chronic medical conditions, particularly with cardiac-related disorders. There is a clear need for clinician-endorsed, evidence-based information on a range of psycho-social and physical health topics for individuals with GA disorders. Such information would assist the patient and their family with treatment decision-making (including weighing up benefits and risks whilst considering their values and life situation), and the self-management of the GA disorder.

As identified by Connors et al (2015), the health information that is most likely to address common unmet information needs in individuals with a GA disorder include: easily
understood medical information about the GA disorder; information about the factors that may affect lifestyle choices; information about risk management; and links to resources that support psycho-social functioning. Benefits of providing patients with this information aims to improve patients’ understanding of the condition and treatment, increase coping, improve medical adherence, and facilitate self-management.

2.1.4 Health Information Sources: Online versus Printed Materials

The availability of information from the Internet has altered the ways that the general population accesses health information and makes health-related decisions (Shuyler & Knight, 2003). It is estimated that of the approximate 16 million Australian who actively use the Internet, almost 80% seek out health information online (Australian Bureau of Statistics, 2014; Nielsen, 2013; McDaid & Park 2011). Data from the US Health Information National Trends Survey that polled over 6,000 adults about their online health activities, levels of trust, and source preference, found that 49.5% of those surveyed preferred going to their physician for specific health information (such as for cancer information). However, only 10.9% of those surveyed had been to their physician first, and 48.6% sought information online in the first instance (Hesse et al., 2005). Similarly, a systematic review conducted by Clarke and colleagues (2015) found that the most common information sources amongst primary care patients were the Internet and clinicians. Online health information seeking has been found to often supplement information from a health professional. For example, Rice (2006) found that 61% of people who looked for health information on the Internet did so in conjunction with a consultation with a physician.
Internet searches for health information are also used to gain increased knowledge of a health condition in order to maximise the visit to the clinician, to ask more relevant questions, and to understand the information and options given in the consultation (Nettleton, Burrows, & O’Malley, 2005). A survey of 285 healthcare professionals in the US found that over 80% reported that patients presented printed information from the Internet at consultations (Podichetty, Booher, Whitfield, & Biscup, 2006). However, it has been reported that due to the increase in individuals looking for web-based health information, the patient-practitioner relationship has changed in both advantageous and less helpful ways (McMullan, 2006; Wald, Dube, & Anthony, 2007). For example, informed patients may communicate better about their medical condition with their clinician and seek care with earlier symptoms (Laing, Hogg, & Winkelman, 2004). Whereas due to variable quality of information on the Internet, consultation time can be taken in discussing misleading or misinterpreted interventions which also result in requests for inappropriate tests or treatment (Ahmad, Hudak, Bercovitz, Hollenberg, & Levinson, 2006; Eysenbach & Köhler, 2002). The reliability of Internet-sourced information is a concern, although it has been reported that patients continue to rely on health professionals to clarify or endorse the information. Members of patient support groups reported a lack of confidence in their ability to discern the quality of health information on the Internet and in printed formats, and relied on authoritative sources such as health professionals to endorse the information (Marshall & Williams, 2006). In a study examining the information needs of individuals with prostate or breast cancer, it was reported that participants were aware of the potential unreliability of online
information, and tended to compare information across sources (Rozmovits & Ziebland, 2004).

Studies exploring the preferred source and type of information materials by patients has indicated that printed materials endorsed by health professionals are highly regarded (Piredda & Marfoli, 2004; Rutten et al., 2005). For individuals with congenital heart defects, the top sources of information rated by importance were physicians (71%), followed by family and friends (58.2%) (Lesch et al., 2014). In rare conditions such as GA disorders, patients need to rely heavily on their treating team for specific and tailored information, in comparison to more commonly known conditions with readily available health education resources. Therefore, there is a clear need for an easily accessible, quality health information resource such as a psycho-education booklet that encompasses medical information, intervention and treatment, and psycho-social impacts and support specific to the GA disorders.
2.2 Written Health Information

2.2.1 Purpose and Objectives of Health Information

Frequently used and preferred health information sources have been found to be one-to-one communication with health professionals supplemented by printed materials (Rutten et al., 2005). Printed health education materials are found for a large range of illnesses and conditions, and particularly for chronic illnesses. These include asthma, dermatology, rheumatoid arthritis, diabetes, chronic obstructive pulmonary disease, over the counter and prescribed medication, pre- and post- surgery, heart disease, and cancers.

A common definition of health education refers to a combination of learning experiences that increase knowledge and skills or influence attitudes in order to facilitate behaviour adaptations that are conducive to better health (World Health Organisation 2013; Bastable, 2006; Falvo, 2010). Studies have shown positive impacts of patient education programs, including written health resources, on patients’ knowledge and recall of health information, health behaviours, and more effective utilisation of health services (Coulter & Ellins, 2006). A review of meta-analyses of patient education in chronic diseases showed that two-thirds of studies that evaluated the efficacy of patient education interventions (including psycho-educational information sessions, one-on-one discussion with a health professional) demonstrated improvements in patients’ quality of life with an impact of between 50-80% (Lagger, Pataky, & Golay, 2010). Patient satisfaction with information provision has also been shown to improve with the use of written information (e.g. leaflets) as a complement to verbal information from health clinicians (Dunnill & Pounder, 2004). Printed health information is important in patient education about their
The Development and Evaluation of a Psycho-education Booklet for Genetic Aortic Disorders

illness, impact, treatment, options, and lifestyle management, and in particular for GA disorders, where easily accessible, clinician-endorsed health education resources are lacking.

There has been some concern that patient education material can increase anxiety in patients, especially considering that patients prefer to receive both good or bad information about their medical condition (Jenkins, Fallowfield, & Saul, 2001). However research indicates that well-designed communication/decision aid resources increase knowledge, reduce decisional conflict, and increase participation in decision making, without increasing anxiety (O’Connor et al, 1999). A study on pre-operative education for open cardiac surgery patients by Asilioglu and Celik (2004) showed no significant difference in anxiety as measured by the Spielberger State-Trait Anxiety Inventory, between those that received preoperative education and those that did not. Various studies in other areas have also shown no adverse psychological distress from exposure to health education such as an information sheet for acute chest pain (Arnold, Goodacre, Bath, & Price, 2009), cardiovascular surgery patients (Yıldız, Gürkan, Gür, Ünsal, Göktas, & Özen, 2014), and cancer (O’Brien, Whelan, Villasis-Keever, Gafni, Charles, Roberts, et al., 2009).

It is commonly found that within health systems, patients are relatively passive recipients rather than being actively engaged in their own care (Nolte & McKee, 2008). With chronic illnesses, long-term self-management of the condition and associated issues is essential to mitigate further medical complications. This can be enhanced with effective
The Development and Evaluation of a Psycho-education Booklet for Genetic Aortic Disorders

health information resources. Written patient education material is therefore important as an adjunct information resource in clinician-patient communication and can play a part in optimising patient care. As discussed earlier in this chapter, given the prevalence of chronic illnesses and evidence of unmet information needs of people affected by chronic conditions, effective means of health information delivery and quality of the information is required, particularly for those affected by a GA disorder where available information can be limited and is not specific for the Australian audience.

Based on the literature, the outcomes of printed health education/information resources would be:

- To help educate the individual affected by the illness, their partner, family members, and support persons
- To increase knowledge of the illness, treatment and lifestyle management
- To promote adherence to treatment
- To assist with possible psycho-social issues and coping

2.2.2 Developing Written Health Information

Learning and educational theories and models

Theories of learning and behaviour change can be applied to how health education materials are developed and disseminated. The development of patient education resources must take learning theories and models into consideration in order to create effective educational tools that enhance information gain and healthful behaviour change.
However, no single theory can explain the complexities of human learning and behaviour, although these models can help guide the development of effective health promotion strategies (Commodore-Mensah & Himmelfarb, 2012). De Lapp (2003) noted that the main theories and models of behaviour change utilised and tested in health include the Health Belief Model, Self-Efficacy Theory, and Stages of Change Model. An overview of these commonly used learning theories in relation to health behaviours will be given here.

The **Health Belief Model** (HBM), first developed by Hochbaum, Rosenstock, and Kegels in the 1950’s was developed to explain the lack of uptake of a free tuberculosis health screening program. Subsequent adaptations of the HBM (e.g. Rosenstock, 1966; Rosenstock, Strecher, & Becker, 1988) have been used to describe that an individual’s belief in a personal threat together with the belief in the effectiveness of the proposed behaviour will predict the likelihood of behaviour change. This model has been applied in a variety of short- and long-term health behaviours, for example, exercise programs, and treatment adherence particularly in persons with a chronic illness (Jones, Smith, & Llewellyn, 2014).

The HBM consists of four main constructs relating to the perceived threat and net benefits which account for a person’s readiness for behaviour change. The constructs are: *susceptibility* (beliefs about the chances of getting a condition), *severity* (beliefs about how serious the condition is and the related consequences of the condition), *benefits* (the beliefs about the effectiveness of taking a particular course of action to reduce risk or
seriousness of the impact), and barriers (the beliefs about negative consequences of following the course of action). Additional concepts, cues to action, activates readiness and cues behaviours through internal or external means, and self-efficacy, is the person’s confidence in their ability to perform an action successfully.

A meta-analysis of the effectiveness of using the HMB to predict health-related behaviour conducted by Carpenter (2010), found that perceived barriers and perceived benefits were the strongest predictors of behaviour. Whereas perceived severity was a weak predictor, and perceived susceptibility was not associated with behaviour change. These findings were supported by Jones, Smith, and Llewellyn’s (2014) meta-analysis of studies using the HBM as the basis for intervention for adherence to medical treatment. Taking into consideration the findings of the meta-analyses as described above, it would be anticipated that behaviours related to severity and susceptibility constructs may require alternate strategies to facilitate appropriate behaviour change.

Nonetheless, HMB can be used to inform strategies to modify beliefs that lead to, or negatively influence, healthful behaviour changes (Rimer & Glanz, 2005). For example, in a psycho-education booklet for GA disorders, the following information can be included to address each component:

- Perceived susceptibility: informing about risk based on typical characteristics or behaviours (e.g. increased risk with high impact sports), and heightening perceived susceptibility if belief is incorrectly low;
• **Perceived severity**: specify and clarify consequences of the risks of the condition (e.g. provide risk information for possible complications and consequences of cigarette smoking);

• **Perceived benefits**: define the action to take, clarify the expected positive consequences (e.g. provide a sample exercise plan with anticipated benefits);

• **Perceived barriers**: identify and reduce barriers through information and support (e.g. provide information and support relating to family genetic testing);

• **Cues to action**: provide “how to” information (e.g. provide an appointment reminder system);

• **Self-efficacy**: provide guidance in performing the desired action (e.g. provide opportunities to document progressive goal-setting based on a personalised treatment plan).

*Self-efficacy theory* refers to an individual’s confidence or belief in their ability to perform a given behaviour. Self-efficacy is specific to the context of the task, meaning that self-efficacy can increase or decrease based on the specific task. An individual can be assisted to modify behaviours by increasing their awareness of skills that can control health events, improving decision-making skills, and the use of social support systems (Bandura, 1995). This translates into health education strategies such as skills mastery and modelling, together with observation and reinforcement which are based on social learning theory. *Social learning theory* first described in clinical psychology by Rotter (1975) and Bandura (1977) explains that learning occurs through the imitation or modelling of behaviours of others and observation, with positive or negative
reinforcements maintaining or decreasing the behaviours. In other words, there is a reciprocal relationship between the individual and the social environment which includes family members, friends, health professionals, colleagues, and others (Rimer & Glanz, 2005). As behaviours change, the social environment and the individual changes in a reciprocal way, therefore changes also impact on health. Consequently, feedback and practical help from others influences health behaviours (Rimer & Glanz, 2005; Israel & Schurman, 1990).

Social cognitive theory incorporates self-efficacy and social learning theory to explain health behaviour change. The three main factors include: self-efficacy, goals, and outcome expectancies (Conner & Norman, 2005). Individuals with a strong sense of self-efficacy are more able to change their behaviours despite obstacles, in comparison to individuals with a poorer sense of self-efficacy, who tend to believe that they have low control over their health behaviour (external locus of control) and therefore feel unable or unmotivated to persist through challenges (Conner & Norman, 2005; Bandura, 1995). Social cognitive theory emphasises the importance of self-efficacy in behaviour change. Strategies that influence behaviour change can therefore include specifying the desired change, breaking down behaviours into small, measurable steps to ensure success, and recognising and celebrating small successes (Rimer & Glanz, 2005). These strategies are pertinent to an information booklet on GA disorders as major lifestyle changes may be required to prevent complications or maintain health. For example, reducing certain physical activities, avoidance of contact sports, or cessation of cigarette smoking. Smaller, specific steps towards change are more likely to facilitate healthful behaviours,
together with offering credible role models who perform the targeted behaviour (e.g. GA disorders support group or individual with a GA disorder who the patient can contact for support). Attention must also be paid to the individual’s social environment, therefore educating and offering resources for the patient’s support person such as their partner or family members is also important (Rimer & Glanz, 2005). A psycho-education booklet that explains GA disorders, possible genetic risks, likely management options, and psycho-social impact of the condition can also be used as an information and communication resource for the support persons of the individuals with a GA disorder.

The *Stages of Change or Transtheoretical model* developed by Prochaska and DiClemente (1985) describes stages that an individual may go through in achieving behaviour change: contemplation, commitment to change, initiation of change, and maintenance. Applying the model to health behaviour change (Rimer & Glanz, 2005), the stages include: pre-contemplation (the problem is not recognised or acknowledged, there is no intention of taking action within the next six months); contemplation (the problem is recognised, there is an intention to take action in the next six months); preparation (some action to resolve the problem has been taken, action is intended within the next 30 days and some behavioural steps towards this has been initiated); action (strategies initiated; behaviour has changed for less than six months); and maintenance (the behaviour change has been incorporated into the lifestyle; behaviour has changed for greater than six months) (Rimer & Glanz, 2005). The transtheoretical model allows the identification of an individual’s motivational and information needs and therefore tailoring of the intervention. The transtheoretical model has been applied across a spectrum of health
issues in chronic illnesses including treatment adherence (Konkle-Parker, 2001), diabetes (Lin & Wang, 2013), medication adherence (Johnson, Driskell, Johnson, Dyment, Prochaska, Prochaska, & Bourne, 2006), and self-management and peer support (MacPherson, Joseph, & Sullivan, 2004).

Given that the booklet in the current study is intended for individuals newly diagnosed with a GA disorder, the earlier stages in the model (i.e. pre-contemplation, contemplation, and preparation stages) are more likely to be relevant to this population. The aims of information provision would therefore be to increase awareness of need for behaviour change (e.g. lifestyle changes), give information about risks and benefits of changes (e.g. risk of worsening versus prevention of aneurysm growth), provide encouragement and information about making plans for change (e.g. where to seek support), and help set gradual goals (e.g. tailor an individual plan for helpful behavioural changes).

It is important that health education/information resources are based on well-established and validated learning theories or health models. Jones, Smith, & Llewellyn (2014) emphasise that health professions need to have clear rationale on their use of theories or health models in their educational activities, interactions with patients, and health promotion programs. However, reporting of the theoretical basis of written health information is rare. In a systematic review of experimental and quasi-experimental studies of cardiovascular health education interventions (such as written material, audio-visual program, or one to one verbal education with a health professional), Commodore-Mensah and Himmelfarb (2012) found that of 25 studies meeting the search criteria, only
one quarter stated the theoretical framework or educational principles that were used to guide the framework of their intervention.

Rankin, Stallings, and London (2005) suggested that an eclectic approach to patient education would best serve the majority of patients. This reflects the approach taken in the development of the psycho-educational booklet for GA disorders as described in this thesis, as it aims to cater to an audience with a range of demographic and personal characteristics, with the commonality of a recent GA disorder diagnosis.
2.3 Evaluation of Written Health Information

2.3.1 Outcomes of Evaluation Studies

There are many evaluation studies of written health information resources such as decision-aids and medication information for patients that show a variety of benefits to patients. However, evaluation studies of written health information for individuals with a chronic illness that specifically examine outcomes such as knowledge gain and retention, and satisfaction with the information or resource, are limited. Historically, it has been reported that only 30% of 183 pamphlets reviewed from within the state of New South Wales in Australia had been evaluated, and only 1.7% had undergone randomised controlled trials (Paul, Redman, & Sanson-Fisher, 1998). More recent information however, is unavailable. At present, and to our knowledge, there have been no formal evaluation of patient educational materials targeting GA disorders.

However, from the available literature on the evaluation of a range of written health resources for chronic illnesses, benefits of health education materials such as leaflets and booklets have been indicated. In particular, patients’ knowledge of their medical condition has generally been shown to increase. An information booklet on stroke (CareFile), which was purpose-designed and individualised for each patient by adding or omitting information, was evaluated for its impact on patients’ level of knowledge and satisfaction with the information (Lowe, Sharma, & Leathley, 2007). The study utilised a pre- and post-test design of 100 inpatients with a primary diagnosis of stroke (n=50 randomised into intervention and control groups). A significant increase in knowledge and awareness of the risk factors of stroke with patients who had received the CareFile
booklet was reported. However, a comparison between groups with/without verbal discussion on the booklet did not show any significant knowledge gain. Knowledge gain was also found in a RCT examining the impact of the addition of commercially produced written information to generic hospital information for elective surgical patients (Sheard, & Garrud, 2006). It was found that those who received additional health information at pre-admission demonstrated an increase in knowledge pre-operatively. Similarly, in an older study by Maggs et al. (1996), a purpose-designed booklet that contained information about arthritis and related matters, including exercising and activities of daily living demonstrated a significant increase in knowledge post-intervention.

However, in a RCT conducted by Venmans, Gorter, Baard, Rutten, and Hak (2007), there was no significant increase in knowledge found between primary care patients with type II diabetes who received a leaflet about common infections (n=100) in comparison to patients who did not receive the leaflet (n=100). Explanations for the lack of effect from the authors was that the material may have been too technical for the target population, that the questionnaire used was not pilot tested, and that the information was not tailored to the individual. This highlights the importance of conducting a pilot evaluation of patient health information, particularly if custom-designed questionnaires or other evaluation tools are used. Lancaster, Dodd, and Williamson (2004) suggested that pilot testing will ensure that data collection tools such as questionnaires are appropriate, comprehensible, and are presented in a consistent way.
As discussed earlier in this chapter, health literacy has been recognised as an ongoing concern amongst the general population, and given the high prevalence of chronic illnesses, it can be assumed that any patient population will also contain a subset of patients who do not have sufficient skills to benefit from the provided information. It is therefore essential to consider the resource’s usefulness for those with lower health literacy in order to determine the material’s target audience, and whether the resource should be suitably modified or if alternate information tools should be offered. This issue was highlighted in a survey study of 363 participants with rheumatoid arthritis given a written information booklet with or without an additional graphical mind map of the information (Walker, Adebajo, Heslop, Hill, Firth, Bishop, & Helliwell, 2007). The findings showed significant knowledge gain overall in both groups as measured using the Knowledge Scale Questionnaire, with no significant difference between groups. However, an important finding with this study was that 15% of the participants assessed using the Rapid Estimate of Adult Literacy in Medicine (REALM), a reading recognition test of medical terminology, were found to have functional illiteracy. The diagrammatic form of the health information was not found to have assisted those with low functional literacy to gain any more knowledge in comparison to only having the written information. It is also important to note that, patients with high literacy levels had significantly lower levels of anxiety and depression. A previous study by Barlow, Pennington, and Bishop (1997) using the same written information without the mind map on rheumatoid arthritis as in the Walker et al. (2007) study, also showed a significant increase in knowledge with exposure to the booklet. In the follow up study (Barlow and Wright, 1998), there was no increase in anxiety found in the intervention group at three
weeks, and the knowledge gain was retained at six months. These studies emphasise the importance of specifically examining why some patients gain little or no knowledge gain, with the growing literature showing that low health literacy is likely to be a major influencing factor. Further research into the effects of low literacy versus higher literacy on psychological factors such as anxiety and depression is also needed for chronic illness populations, and specifically, in those with a GA disorder.

There is very limited literature on the impact of written health information on areas apart from knowledge, such as the impact on self-management and psycho-social aspects. However, in a meta-analysis of the effect of educational material in diabetes, Cooper, Booth, Fear, and Gill (2001) found the greatest effect for knowledge, moderate effect for physical impact, and smallest effects for self-care activities and psychological impact. The authors noted that the interventions were not well described, that outcome tools were not validated, and only short-term benefits of up to six months were assessed. There is clearly a need for evaluation studies to be conducted on written health information encompassing a greater range of outcome measures and on its longitudinal effect.

Another potential issue concerning the evaluation of written health information is that the resources are most commonly evaluated for readability, rather than on quality of the content and patient satisfaction with the information. As discussed above, the readability level of the majority of available health resources is higher than generally recommended. However, important findings about readability and content were reported in Rhee, Von Feldt, Schumacher, & Merkel’s (2013) readability and suitability assessment of patient
education materials for several rheumatic conditions. Of the twenty-three printed resources evaluated, only one met the recommended reading level of sixth-grade reading level as measured by the Flesch-Kincaid readability test, and five of the resources received superior suitability scores, with the rest receiving only adequate suitability scores as measured by the Suitability Assessment of Materials (SAM). The findings highlight that readability and content should both be evaluated, given that though an information resource may meet an appropriate standard for readability, the content may not be adequate.

In one of few studies specifically assessing the quality of the content of a leaflet, it was found that the information in leaflets on genetic testing for patients and families (n=50), was lacking in a variety of topics. The topics that were lacking included: post-test information; patient rights; and information about shared decision-making. In addition, the leaflet was unbalanced, containing more information about benefits of genetic testing in comparison to its risks (Lewis, Mehta, Kent, Skirton, & Coviello, 2007). This finding highlights that much more attention needs to be paid to assessing the quality of the content in written health information and whether the information meets the information needs of its intended readers. The development of the current study’s psycho-education booklet is based on a comprehensive study that described unmet information needs of individuals with a GA disorder (Connors et al., 2015) complemented by consultation with health specialists, in order to determine the type of content that would best meet the information needs of persons affected by a GA disorder. The pilot evaluation of the booklet will also focus on assessing the appropriateness and value of the content. Finally,
the booklet development and assessment follows the comprehensive and widely used C.R.E.D.I.B.L.E. criteria (O’Connor et al., 2003), developed as part of the Cochrane Systematic Review of Patient Decision Aids.

Limitations common to the evaluation studies reviewed include the lack of description of the development process of the material, validation of the evaluation tools used, and large non-response rates. Further, Sheard and Garrud (2006) commented that there is insufficient evaluation of patient information resources that determine whether they increase patient satisfaction and post-treatment well-being.

Overall however, knowledge has commonly been found to be improved through the use of printed educational material. A secondary outcome commonly reported in studies was patients’ views about the material. There was general consensus across studies that patients found leaflets or booklets useful and valued the resource.

2.3.2 Tools and Guidelines for the Evaluation of Written Materials

A number of tools have been developed to evaluate the quality of written health materials, particularly for patient decision aids. Decision aids are used to inform patients of treatment options and the risks and benefits associated with the options, and to assist in evaluating and sharing in treatment decisions. A short instrument, DISCERN, was developed by Charnock, Shepperd, Needham and Gann (1999) to assist with assessing the quality of written consumer health information on treatment choices. This instrument was developed to be used on resources such as decision aids. The assessment tool
consists of fifteen questions with a five-point scale ranging from No to Yes, and an overall quality rating. The questions involve the domains of reliability of the publication, and specific details of the information about treatment choices. DISCERN was not designed to assess the quality or accuracy of the content, rather, it was developed to assess what information is provided.

A tool to measure content, or suitability, of printed health-related educational resources is the Suitability Assessment of Materials (SAM; Doak et al., 1996). The SAM consists of six criteria which the resource is measured against: content (e.g. behavioural instructions) literacy demand (e.g. common words are used); graphics (e.g. simple line drawings); layout and typography (e.g. at least 12pt font size); learning stimulation and motivation (e.g. subdivision of complex topics); and cultural appropriateness (e.g. appropriate and positive representation of the culture through use of images and examples). The SAM has been used in various studies of written health information in chronic illnesses, for example, prostate cancer (Weintraub, Maliski, Fink, Choe, & Litwin, 2004), chronic kidney disease (Tuot, Davis, Velasquez, Banerjee, & Powe, 2013), and stroke (Hoffmann & Ladner, 2012). Luk and Aslani’s (2011) review of 64 articles that evaluated written health information and written medicine information identified that the most common tool for readability was the SMOG, and for presentation was the SAM.

The most current assessment tool is the IPDASi - International Patient Decision Aid Standards instrument (Elwyn et al., 2009) which was developed to provide quantitative assessments of patient decision support interventions and technologies in terms of their
quality at item, dimension, and global levels. The assessment aims to provide feedback across ten broad dimensions about the content, development, implementation, and evaluation of the decision aid.

The Australian National Health and Medical Research Council developed guidelines on presenting evidence-based health information in consumer publications based on a review and summary of the literature on consumer needs assessment and preference, evaluation, and process studies (National Health and Medical Research Council Australia, 2000; Barratt, 2000). The recommendations relate to considering consumers’ health information needs and preferences, evaluations of consumer health information, and the process of developing information materials.

The tools described above have been shown to be valid and useful for health information resources such as decision-aids for a spectrum of patient populations. However, the booklet in the current study is not intended as a decision-aid, and the pilot evaluation of the booklet aims to be comprehensive, in-depth, and encompass multiple aspects of suitability, content, and readability, therefore assessment tools designed for quick assessments are not suitable. Hence, the current study will utilise custom-developed questionnaires to assess multiple factors of the booklet, guided by and expanded on the tools and guidelines as described above. Systematic evaluation tools as described above would be appropriate for the RCT phase of the evaluation of the booklet.
Summary

Individuals affected by chronic illnesses, including GA disorders, generally want as much information as possible, and in particular, prefer to have information specific to their condition, stage of disease, treatment options, risks, benefits and likely outcomes of treatment options, and practical information about lifestyle and psycho-social changes. This information should be evidence-based, easily accessible and endorsed by health professionals. Written health information must take into consideration the health literacy demands on the target audience, and the design elements that influence the readability and suitability of the resource. The process of developing the resource must also involve a systematic assessment of its acceptability, safety, usefulness and potential impact on the intended audience. The current study aims to fill the identified gap in information resources for individuals diagnosed with a GA disorder by developing and pilot-testing a printed psycho-education booklet based on evidence base, health professional, and patient input. The aims of the current study are presented below.
2.4 Aims of the Research

The aims of the study were to:

1. Develop an evidence-based psycho-education booklet for patients recently diagnosed with a GA disorder

2. To evaluate the booklet for clarity, usefulness, and acceptability among:
   
a) Patients diagnosed with a GA disorder, and
   
b) Clinicians/health professionals specialising in GA disorders

The rigorous process of the booklet development will be presented in the next chapter.

The method and results of the evaluation phase, the overall discussion, clinical implications, limitations, and conclusions will follow in subsequent chapters.
Chapter 3: The Development of a Psycho-Education Booklet for Individuals with a Genetic Aortic Disorder

The content and format of the booklet was based on an extensive literature review and reiterative consultations and feedback from a team of clinical experts in GA disorders, experts in health communication and patient education material development, and patients affected by GA disorders. The steps in the development of the booklet are described in this chapter.

3.1 Development and Evaluation of the Booklet

3.1.1 Overall process – Introduction

It is known that specific attention needs to be paid to various factors that affect the quality of the resource throughout its development. These concerns include the resource’s quality, reliability, readability, usefulness, and accessibility (Coulter & Ellins, 2006). It is also important that the end users of the resource, the patients, be involved in the development and evaluation of information materials (Coulter, Entwistle, & Gilbert, 1999).

Checklists and guidelines have been developed to address issues as described above and in Chapter 2 (Sections 2.2 and 2.3). For example, Coulter et al. (1999) suggested the following process for the development of patient information materials, which was used to guide the development of the current resource:
• Involve a wide range of clinical experts

• Be specific about the purpose of the information and target audience

• Consider the information needs of minority groups

• Review the clinical research evidence and use systematic reviews where possible

• Plan how the materials can be used within a wider program promoting shared decision making

• Consider cost and feasibility of distribution and updating when choosing media

• Develop a strategy for distribution

• Evaluate the materials and their use

• Make arrangements for periodic review and updating

• Publicise the availability of the information materials

The development of the booklet was also guided by the commonly used and supported Ottawa guidelines (O’Connor & Jacobsen, 2003) for decision aid development and evaluation. The guidelines consist of seven main steps: assess need; assess feasibility; define the objectives; identify the framework; select the methods of decision support; select the designs and measures for the evaluation; plan dissemination. Although the GA disorders booklet was not intended as a decision aid, the Ottawa guidelines provided general guiding principles that were relevant for the development of a psycho-education resource.

The following sections describe specific aspects of the development and evaluation of the booklet in detail.
3.1.1.1 Assess need
As discussed in Chapter 2.1, there is clearly a need for written health information to support individuals diagnosed with a chronic illness, and with GA disorders in particular. A mixed method study by Connors et al. (2015) reported that participants with GA disorders preferred to receive information from experts in the area of GA disorders as they were trusted sources. Also that informational resources are needed to aid in the understanding of the disorder and treatment options and to make the treatment process easier to navigate. Within the current study, discussions with clinicians involved in the care of individuals with GA disorders or associated chronic conditions strongly endorsed the need for an easily accessible, evidence-based resource for their patients.

3.1.1.2 Assess feasibility
Discussions with relevant clinicians and health education specialists showed that a psycho-education booklet would provide an easily accessible resource. There were no concerns about any barriers to the dissemination of a psycho-education booklet to patients.

3.1.1.3 Define the objectives of the booklet
The common issues identified in the literature relating to patients’ information needs after a diagnosis of a medical condition include the prevalence and aetiology of the condition, its consequences, recovery timeframe, and possible outcomes (Clarke et al., 2015; Coulter & Ellins, 2006). There is a clear preference for receiving relevant information soon after the diagnosis, with more specific information wanted as the need emerges over
time, such as with issues of self-care and long-term prognosis in cancer patients (Rutten et al., 2005; Piredda & Marfoli, 2004). These identified information needs were reflected in the objectives of the current booklet. The objectives of the booklet were defined as follows:

- To facilitate information delivery in commonly identified issues including:
  (medical) knowledge about the diagnosis and treatment options, addressing common concerns, information about self-care issues, lifestyle changes, and other frequently asked questions;

- To facilitate communication between patient and clinician about GA disorders diagnosis and treatment, to assist with treatment decision-making, and in making collaborative treatment plans;

- To have available an easily accessible psycho-education resource that can be used and adapted over time with further information and developing treatment plans.

To aid in meeting these objectives, the theoretical principles and models that guided the framework and content of the booklet included the Health Belief Model, Social cognitive theory, and Transtheoretical model as described in detail in Chapter 2 (Section 2.2.2). This integrative approach was considered to be most appropriate and relevant to the target audience of the booklet given the likely range of demographic, personal, medical characteristics, and information needs of individuals recently diagnosed with a GA disorder.
3.1.1.4 Select the design and measures to evaluate the booklet

A mainly quantitative methodology using questionnaires, supplemented with a qualitative component involving semi-structured interviews, was used to evaluate the booklet. The questionnaires consisted of purpose-designed, and commonly used standardised research and clinical measures. A purpose-designed scale was developed to gather participants’ feedback about the content, format, acceptability, and readability of the booklet. Standardised scales used included the Depression, Anxiety and Stress Scale (DASS21) which measures psychological well-being, and a modified Information and Decision Making Preference Scale which measures the level of information preferred by the patient about their condition and treatment options. Demographic and clinical information about participants was also obtained. Opportunities for participants to give qualitative feedback using open-ended questions were given within the study questionnaires.

An important and very informative aspect of the evaluation was the 20-25 minute semi-structured interviews with an additional sample of patients. These interviews provided in-depth feedback and comments about the booklet and elicited responses, suggestions, opinions, and views for specific components of the resource.

3.1.1.5 Plan dissemination

As part of this study, the booklet was intended for use within a specific service (cardiology) provided by a member of the research team (GA disorders specialist). As GA disorders are considered a specialist area, only a relatively small number of clinicians have expertise in this area and patients are referred or seek consultations with these experts from a broad geographical area around Australia. It is hoped that an electronic
version of the booklet would facilitate the accessibility of the booklet and enable a wider
distribution via services with clinicians that specialise in GA disorders.

3.2 The development of the booklet

At the first stage of the booklet development, a comprehensive literature review was
conducted, and other relevant resources such as any type of information resources (e.g.
printed, video, online) relevant to GA disorders was reviewed. An initial draft of the
booklet was developed based on the literature findings, relevant theoretical frameworks
relating to health education, and guidelines as described in Chapter 2 and earlier in this
chapter. The overall development of the booklet followed an iterative process of drafting
and refining the booklet. The iterative review process involved discussion, re-drafting and
review by a working party, which comprised of the research team and clinicians, who
refined the booklet to ensure that its quality reflected the most up-to-date research
evidence and best clinical practice for individuals diagnosed with GA disorders.

In terms of the content of the health information resource, Entwistle, Sheldon, Sowden,
and Watt (1998) emphasised the importance of including up-to-date information based on
evaluated research, the use of appropriate language, terminology, style, and tone, and
clearly stating the purpose of the material. These elements were incorporated into the
draft as described below.
Language

Coulter, Entwistle, and Gilbert (1999) suggested using non-alarmist, non-patronising language in active rather than passive voice. Coulter & Ellins (2006) also support the use of the active voice, the use of common words, and context placements. The study booklet focused on the use of layman’s terms and the use of diagrams to explain medical terms and complex medical conditions and treatments, as well as using straightforward language with a sensitive tone. Emotive and overly dramatic words, phrases and tone were avoided.

An excerpt from the booklet’s section on Myths and Misconceptions about life expectancy is given to illustrate the tone and language used in the booklet:

Myth: Life expectancy is poor

Twenty years ago, the outlook for patients with a GA disorder was more limited. In the current era, with active monitoring, medical, and surgical treatment, the life expectancy for individuals with a GA disorder nears that of the general population. An active, full, and long life can be expected.

Framing

The way that risks and probabilities are framed and presented is an important issue to consider. The information can be presented in various forms such as text, numeric, and graphic form, and in combination, can increase patients’ understanding and responsiveness to the risk information (Jones & Iverson, 2005). It is also known that framing of data can influence the perception of risk, for example, presentation of relative
risk (e.g. 50% increase of being affected by a condition) can have more of a negative impact than absolute risk (e.g. 1 in 20) (Edwards, Elwyn, Covey, Matthews, & Pill, 2001). Descriptors such as “common”, “uncommon” and “rare” can be interpreted incorrectly as an overestimation of the probability of adverse effects in comparison to when numerical information is used (Büchter, Fechtelpeter, Knelangen, Ehrlich, & Waltering, 2014). Descriptors can also be interpreted differently by different people, for example, ‘infrequent’, ‘vast majority’, ‘atypical’, has been found to have different meanings and estimates to different individuals (Nakao & Axelrod, 1983). It is therefore better to use number specifications rather than descriptor words, as well as absolute rather than relative risk.

A positive frame was consistently utilised in the study booklet with avoidance of descriptor and easily misinterpreted words. Risks and probabilities were presented in easily understandable quantified terms. An example from the booklet: “The condition occurs in approximately 1 out of 5,000 people and affects both males and females of any race or ethnic group.”

Specific elements
To encourage shared management between patients and their doctors, it has been shown useful to include sections such as “Your Choice” and “Test Results” (Kennedy & Rogers, 2002). Similarly, the current study booklet included the sections ‘My Questions’, ‘Treatment Plan and Recommendations’ and ‘Test Results’ to give opportunities for
patients to document personal information, to note their own comments, and to facilitate informed decision-making and open communication between patient and clinician.

Doak, Doak and Root (1996) in their suitability assessment of material for low literacy readers suggested that important elements of graphics are those that visually carry content. Connors et al. (2015) specifically suggested that printed information in the context of GA disorders contain the following elements: pictures and information about the heart and related systems, a list of signs and symptoms, a risk management plan, links and contacts, and that terminology be explained. In support of these findings, the text of the booklet was supported by clear, simple graphics. Some of the graphics were of real situations, e.g. X-rays, others were simplified representations, e.g. aortic grafts. Focus boxes were used to highlight important information, such as the purpose of medication, suitable activities, or stress management tips. An emergency plan with a list of signs and symptoms, instructions, and emergency contact information was also included. A glossary of medical terms used in the booklet and a section providing links for further information on GA disorders, information networks, and support groups were included in the booklet.

Coulter, Entwistle, & Gilbert (1999) suggested the inclusion of checklists and questions to ask the clinician, and to address common concerns and misconceptions. A review of the use and effectiveness of question prompt lists (QPL) by Sansoni, Grootemaat, and Duncan (2015) found that the communication between patients and clinicians improved with the use of QPLs. Questions asked by patients are increased with QPLs about
The Development and Evaluation of a Psycho-education Booklet for Genetic Aortic Disorders

prognosis, diagnosis, and treatment issues (Brown, Butow, Dunn, & Tattersall, 2001; Bruera, Sweeney, Willey, Palmer, Tolley, Rosales, & Ripamonti, 2003; Smets, Van Heijl, Van Wijngaarden, Henselmans, & Van Berge Henegouwen, 2012). Effective QPLs contain questions about treatment, test options, the risks and benefits, and the likelihood of the risks and benefits occurring. A specific page in the current booklet provided a checklist of questions as suggestions to patients to discuss with their clinician. This was entitled “*Questions to ask your physician*”. In addition, a page titled “*Myths and Misconceptions*” was developed to address common concerns and correct common erroneous beliefs.

Doak, Doak, & Root (1996) recommend that the layout and typography include content chunking. The contents of the current study booklet was grouped into sections to facilitate a logical flow of information and reflect the process of a typical process from diagnosis to treatment planning and longer-term management and including important well-being oriented elements such as emotional adjustment and coping support. The content was arranged into three main sections:

1. **Factual information**: medical and other information about GA disorders and associated conditions, treatment, and emergency plan;

2. **Person and lifestyle related information**: lifestyle issues, frequently asked questions, myths and misconceptions, and psychological and well-being aspects;

3. **Practical and support-oriented information**: a question checklist, evolving treatment plan and recommendations, appointments, test results, further information and resources, and glossary
The full content list is presented in Table 3.1 and some exemplar pages are given in Figures 3.1-3.5.

The final booklet is provided in the Appendices (Appendix J).

Table 3.1 Booklet Content List

<table>
<thead>
<tr>
<th>Contents</th>
<th>Page</th>
</tr>
</thead>
<tbody>
<tr>
<td>How This Booklet Can Help You</td>
<td>4</td>
</tr>
<tr>
<td>Josh’s Story</td>
<td>5</td>
</tr>
<tr>
<td>About Genetic Aortic Disorders</td>
<td>6-7</td>
</tr>
<tr>
<td>What is an Aortic Aneurysm?</td>
<td>8</td>
</tr>
<tr>
<td>What is an Aortic Dissection?</td>
<td>9</td>
</tr>
<tr>
<td>How is an Aortic Aneurysm Diagnosed?</td>
<td>10</td>
</tr>
<tr>
<td>Treatment: What to expect</td>
<td>11</td>
</tr>
<tr>
<td>Surgical Repair</td>
<td>12</td>
</tr>
<tr>
<td>In an Emergency</td>
<td>13</td>
</tr>
<tr>
<td>Do I need to make changes to my lifestyle?</td>
<td>14-15</td>
</tr>
<tr>
<td>Other Frequently Asked Questions</td>
<td>16</td>
</tr>
<tr>
<td>Myths and Misconceptions</td>
<td>17</td>
</tr>
<tr>
<td>Coping with the emotional ups and downs</td>
<td>18</td>
</tr>
<tr>
<td>My Thoughts and Feelings</td>
<td>19</td>
</tr>
<tr>
<td>Questions to Ask Your Physician</td>
<td>20</td>
</tr>
<tr>
<td>My Questions</td>
<td>21</td>
</tr>
<tr>
<td>My Treatment Plan and Recommendations</td>
<td>22</td>
</tr>
<tr>
<td>Appointments</td>
<td>23</td>
</tr>
<tr>
<td>Test Results</td>
<td>24</td>
</tr>
<tr>
<td>More Information and Resources</td>
<td>25</td>
</tr>
<tr>
<td>Glossary of Terms</td>
<td>26</td>
</tr>
<tr>
<td>Notes</td>
<td>27</td>
</tr>
</tbody>
</table>
The Development and Evaluation of a Psycho-education Booklet for Genetic Aortic Disorders

Figure 3.1 Booklet -Front Page

Figure 3.2 Booklet -Patient’s Story

Josh’s Story

“I am 26 and was about to travel in Asia with my girlfriend. My family doctor said he heard a heart murmur and sent me for tests. Next thing I know, I’m seeing a heart specialist who says I have a serious problem with my aorta and I will need surgery soon. Then he said I can’t do weight training anymore. Where did this come from? Why me? I haven’t done anything to anybody…”

“My world was turned upside down for a while, but I was determined to find out as much information as I could about what I needed to do to recover after the surgery. I also talked to my doctor and my specialist about what I needed to do to stay healthy in the long term. Thinking about going on the trip with my girlfriend was the thing that kept me going on the down days. Guess what? We’ve just returned from an awesome 6 month trip!!!”
The Development and Evaluation of a Psycho-education Booklet for Genetic Aortic Disorders

Figure 3.3 Booklet – What is an Aortic Aneurysm?

What is an aortic aneurysm?

An aneurysm is a bulging or ballooning in the wall of an artery, usually in a weak area of the aorta. The pressure of blood inside the artery forces the weak area to balloon outward.

Symptoms of an aortic aneurysm may include chest or back pain or swelling. An aneurysm may rupture, resulting in internal bleeding. These complications can be avoided through detection, surveillance, and surgical repair of the aneurysm.

Modern medical surgery has provided patients and those in a wide variety of high-risk factors, including balloon implants, stenting, and valve repairs and replacements which are performed by a highly-trained surgical team.

Figure 3.4 Booklet – Myths and Misconceptions

Myths and Misconceptions

Vitamins and diet can protect the aorta.

A healthy diet is important for cardiovascular health in everyone, but there is no evidence that dietary supplements or any particular diet can protect the aorta. However, maintaining a healthy weight can help to reduce the risk of an aneurysm and it is important that you have your cholesterol levels checked and treated if too high.

Life expectancy is poor.

Twenty years ago, the outlook for patients with a GA disorder was much limited. In the recent era, with better monitoring, medical, and surgical treatment, the life expectancy for individuals with a GA disorder now resembles that of the general population. As a result, a full and normal life can be expected.

I can’t travel overseas.

There is no restriction on travel, although visiting very high altitudes requires more caution. Individuals with GA disorders have traveled all over the world. Be sure to let your physician know if you plan to travel, so medical records and documents can be organized. If you are going to live overseas, medical contacts for medical check-ups in other countries can be arranged.

Men get aortic aneurysms.

The inheritance of GA disorders affects men and women equally. However, it is true that men are more likely to develop aortic aneurysms than women. The reason for this difference is currently unknown.
The Development and Evaluation of a Psycho-education Booklet for Genetic Aortic Disorders

Figure 3.5 Booklet – Questions to Ask Your Physician

Questions to Ask Your Physician

- What is the diagnosis?
- What are my symptoms?
- What are the risks and benefits of not treating the condition?
- What are the risks and benefits of treating the condition?

Tests:
- How often will I need to have tests?
- Are there any precautions I should take before the test?
- Will it hurt to take the test?

Prognosis:
- What is the likely outcome with this condition?
- What is the best case scenario?
- What is the worst case scenario?

Treatment Information:
- What treatments are available for this condition?
- What is the purpose of taking medication or having surgery?
- What are the possible side effects of the medication?
- What are the potential risks and long-term options?

Lifestyle:
- What is the best way to lose weight?
- What are the benefits of exercise?
- What is the best diet for this condition?

Costs:
- What is the cost of the treatment?
- What is the cost of the surgery?

Information and Support:
- What is the best source of information about this condition?
- Can I talk to someone about how to cope with the stress?
3.3 Rigour and quality of the booklet

Rigour and quality of the booklet is an important aspect of any health education resource. The widely used C.R.E.D.I.B.L.E. criteria (O’Connor & Jacobsen, 2003, p. 327), developed as part of the Cochrane Systematic Review of Patient Decision Aids, were used to guide the development and assessment of the booklet. How each element of the criteria are addressed in the current booklet is documented below.

C - Competently developed:

*Are the essential components that promote quality decision-making included?*

Essential information about treatment and lifestyle decisions were included in the booklet to improve understanding of the condition and promote informed choice about management options.

*Are the credentials of developers included in the booklet or supporting materials?*

Relevant details of the developers were included in the acknowledgement page.

*Is the development process adequate?*

A needs assessment (previous extensive research project on unmet needs, and updated needs review) and review by a panel of experts and users guided the development process.

R - Recently updated:

*Does the developer have an update policy or evidence review process that is continuous*
or at least every two years? It is anticipated that the booklet will be regularly updated to incorporate most up-to-date evidence-based information, in consultation with clinicians and patients.

E - Evidence-based:

Is there a link to an evidence review group or is the process that was used to identify and appraise evidence described?

The extensive literature review used to inform the evidence-based information in the booklet has been described in Chapters 1 and Chapters 2.

Are references to scientific studies or systematic overviews used to support statements describing benefits/harms?

A list of further information resources was provided in the booklet. Patients are directed to follow links to up-to-date clinical and scientific studies and information.

DI - Disclosure of conflicts of Interest:

Is there disclosure of sponsorship and conflict of interest?

There was no sponsorship or known conflict of interest in developing the booklet.

BL - Balanced presentation of options, benefits, and harms:

Is there a balanced presentation of potential harms and benefits?

Information about options, benefits and harms are given in a neutral stance to avoid
perceived bias to any one option. In addition, equal amounts and depth of information was given for each option.

*Do the majority of users find it balanced?*

Perceived balance of information was assessed during the evaluation phase (See Chapter 4 and 5).

**E – the material is Efficacious at improving decision-making:**

*Do evaluations show that the decision aid improves knowledge? Do evaluations show that the decision aid is acceptable to users? Do evaluations show other benefits? Do evaluations show that it was free from adverse effects?*

These aspects were assessed in the evaluation phase (See Chapters 4 and 5).

*Do evaluations include a randomized controlled trial?*

Since it was not in the scope of the current thesis to conduct a randomised controlled trial, the booklet underwent a comprehensive pilot evaluation.

It is acknowledged that the International Patient Decision Aid Standards instrument (IPDASi, Elwyn et al., 2009) based on the IPDAS criteria (Elwyn et al., 2006), provides a way of quantitatively assessing the quality of decision aids at an item, dimension, and global level. However, since the current study booklet was not designed to be a decision aid, many of these criteria were not applicable and a thorough assessment using IPDASi
therefore was not implemented. Nevertheless, the principles of IPDAS/IPDASi influenced the development of the booklet in ensuring its rigour and quality.

**Post-Evaluation Review**

Review meetings among the research group members were held regularly post-survey collection and interview phases, to discuss and implement modifications to the booklet in consideration of the patient and clinician feedback.

**Summary**

The process of the development of the study booklet, a psycho-education booklet on GA disorders, was presented in this chapter. The factors that influence the quality, reliability, readability, usefulness, and accessibility of written information resources were outlined and how these factors related to the current study’s booklet was discussed. The process of ensuring rigour and quality was presented.

The following chapters will present the pilot evaluation of the booklet, the outcomes, discussion, and conclusions.
Chapter 4: Pilot Evaluation of the Booklet

As described in Chapter 3, a psycho-educational booklet for individuals with a Genetic Aortic disorder was developed based on an extensive literature review, discussion with experts in the field of GA disorders and in health education development and evaluation, and updated needs assessment. This chapter presents the process and results of the pilot evaluation of the booklet.

4.1 Aims

The evaluation phase of the study aimed to assess the booklet’s clarity, usefulness, and acceptability among:

a) Patients diagnosed with a GA disorder
b) clinicians/health professionals specialising in GA disorders

4.2 Design

Two participant cohorts – patient group and clinician group - were recruited for the pilot evaluation phase of the study. A mixed methods approach was used to obtain feedback of the booklet. The quantitative component of the evaluation comprised a series of purpose-designed and standardised research and clinical measures, completed by both (patient and clinician) cohorts. The qualitative component of the evaluation involved semi-structured interviews conducted with patients only; with the purpose to obtain in-
depth booklet feedback and to elicit responses, suggestions, and views about specific components of the information resource.

4.3 Participants

Patient Group – Questionnaires and Semi-structured Interviews

The patient group participants were recruited through the Marfan and Aortic Disease Clinic, Royal Prince Alfred Hospital (RPAH), Sydney. The Clinic is a part of the Sydney Heart Centre, an annexe of the RPAH Cardiology Department, and runs fortnightly. The Clinic Director is a specialist in the diagnosis and treatment of individuals with a GA disorder. Patients of the clinic come from metropolitan and regional areas of NSW, as well as interstate.

The inclusion criteria for the patient group included: over 18 years of age, proficiency in English to participate in the study, with a recent or previous diagnosis of a GA disorder. Exclusion criteria included individuals with an untreated psychiatric illness, unable to give informed consent, or insufficient English proficiency to complete questionnaires/interview.

The target sample size for the questionnaires was set at 20 patients. Previous pilots of communication aids (Marshall & Williams, 2006) have shown that approximately 20 participants are sufficient to: a) reach theoretical saturation in qualitative feedback, and b) provide trends on quantitative measures.
Clinician Group - Questionnaires

Participants of the clinician group were specialist clinicians involved in the care of patients with a GA disorder. The exclusion criteria included: clinicians who were not currently involved in the care of patients with a GA disorder. In order to maximise recruitment numbers, the clinician sample was based on a convenience sampling procedure. The field of specialist clinicians with particular expertise in GA disorders is very small and together with a mail back procedure, the recruitment of this group was expected to be limited.

4.4 Procedure

4.4.1 Ethics

Ethical approval was obtained by the Sydney & South West Area Health Service, Human Research Ethics Committee (HREC) with Site Specific approval for the Marfan and Genetic Aortic Disease Clinic, RPAH (Protocol X13-0061, see Appendices A & B).

Ethical approval was ratified by the University of Sydney HREC.

4.4.2 Procedure - Patient Group

Eligible participants were identified by the Clinic Director when attending their medical consultation. The Clinic Director briefly explained the study, and invited patients to participate in the study. Patients interested in participating in the study were provided with a pack containing the study invitation letter (Appendix C), participant information form (Appendix D), consent form (Appendix E), the patient education booklet (Appendix
J), the questionnaire (Appendix F), and a reply-paid envelope. In addition, eligible patients who were involved in a previous research study on coping with GA disorder, and who had indicated an interest in participating in further research were mailed the same study pack.

Those wishing to participate were instructed to sign the consent form, read through the booklet, and then complete the questionnaire. Once completed, they were asked to send the questionnaire together with signed consent form back to the researchers using the pre-paid envelope provided.

The questionnaire included questions on demographic information, psychological wellbeing, and feedback about the format, content, and usability of the booklet. It was estimated that reading the booklet and completing the forms would take approximately 30-40 minutes. Patients not interested in the study were instructed to discard the study pack. As this was an opt-in study, patients who did not respond were not followed up.

All potential participants were informed that their participation was voluntary, that they were free to withdraw at any time without having to give a reason, and non-participation or withdrawal from the study would not affect their care or relationship with medical staff at the Clinic.
4.4.3 Procedure - Clinician group

Eligible clinicians identified by the Clinic Director and the research team were invited to participate in the study via email or letter from the Clinic Director. The email included a Clinician Information Letter (Appendix H) and instructions to access the booklet and anonymous questionnaire online. Those wishing to participate were instructed to click on the provided web link and enter the given password to access the online Clinician Information Letter, study booklet and questionnaire. Before accessing the questionnaire, potential participants were instructed to read the booklet. They were also informed that the survey was anonymous and that they could withdraw at any time by closing the survey window. The questionnaire included items assessing demographic and clinical practice information, as well as feedback regarding the format, content, acceptability and usability of the booklet (Appendix I). It was estimated that reading the booklet and completing the questionnaire would take approximately 20-30 minutes. Alternatively, potential participants were posted a study pack with the anonymous questionnaire and instructed to mail back the completed questionnaire in the replied paid envelope.

4.4.4 Quantitative Measures

*Patient Questionnaires*

The patient questionnaire set included the following standardised and purpose-designed measures, used to characterise the sample and gain both quantitative and qualitative feedback on the booklet. The following questionnaires were included:
Demographic and clinical characteristics: This purpose-designed questionnaire was used to gather information about participant demographics and clinical factors, including their age, relationship status, education, occupation, type of GA disorder, and treatment.

Booklet Feedback Questionnaire: This purpose-designed questionnaire was used to gather participant feedback on the usefulness, design, and acceptability of the booklet. The questionnaire was based on those used in previous patient psycho-education and decision-making tools (e.g. Juraskova et al., 2015; Juraskova, Butow, Bonner et al., 2014; Anderson, Carter, Nattress et al., 2011). The questionnaire consisted of twelve statements, with five response options ranging from “Strongly Disagree” to “Strongly Agree”. Examples of statements include: “The booklet was easy to read”, and “The booklet answered questions about how my condition may affect me”. Opportunities for qualitative comments and feedback were also included.

Information and Decision Making Preferences: This scale was adapted from the Control Preferences Scale (CPS; adapted from Degner, Sloan, & Venkatesh, 1997), and Information Style Questionnaire (ISQ; Cassileth et al., 1980). The control preferences construct is defined as the level of control that an individual prefers in relation to treatment decisions. The ISQ assesses how much medical information is preferred by patients. Two items elicit preferences for patients’ participation in treatment decision-making using the statements ‘I prefer to leave decisions about my medical care and treatment up to my doctor’ and ‘I prefer to participate in decisions about my medical
“care and treatment”. Both ISQ and CPS are commonly used measures in psycho-social studies involving medical samples.

In the current study, based on the CPS and ISQ, a continuous rating scale from 0 (least) to 10 (most) was used. Participants were asked to select on the scale the point that reflected their preference for details about their condition in relation to the management of their condition. The scale consisted of the two options: “Prefer as few details as possible” (0) to “Prefer as many details as possible” (10). Other studies have supported the use of a single option measure (e.g. Davison, Goldenberg, Wiens, & Gleave, 2007).

Depression, Anxiety, and Stress Scale (DASS 21, Lovibond & Lovibond, 1995): The DASS21 was used to assess participant psychological wellbeing, including levels of depression, anxiety, and stress. The DASS21 is a 21-item questionnaire designed to assess an individual’s self-rated level of depression, anxiety and stress. The DASS21 is a widely used self-report instrument. The scale consists of 21 items assessing three subscales, with four-point response options ranging from “did not apply to me at all” (0) to “applied to me very much, or most of the time” (4).

The scale has robust psychometric properties with moderate to high reliability and validity. Cronbach’s alpha for all subscales is greater than or equal to 0.92, indicating high internal consistency of items. Concurrent validity of the Depression scale of the DASS21 with the Beck Depression Inventory (BDI) is 0.77, and validity of the DASS21 Anxiety scale with the Beck Anxiety Inventory (BAI) is 0.84 (Antony et al., 1998).
Previous research in chronic disease populations have utilised the DASS21/DASS42 (e.g. Covic, Cumming, Pallant, et al., 2012; Kaur, Tee, Ariaratnam, Krishnapillai, & China, 2013; Khan, Amatya, Pallant, Rajapaksa, & Brand, 2012).

The DASS21 was selected over alternate measures as it gives information about depression, anxiety, and stress within the one assessment tool. Alternatively, participants would need to complete several separate tools, such as the Beck Depression Inventory and Beck Anxiety Inventory which would be more time-consuming. In addition, the DASS provides norms for the Australian population (Crawford, Cayley, Lovibond, Wilson & Hartley, 2011).

Scores from the items are summed according to one of the three sub-scales. Higher scores in each sub-scale are considered to indicate higher levels of the relevant domain. Sub-scale scores range from 0 (normal functioning) to 34+ (extremely severe distress) depending on the subscale (see Table 4.1). The scale demonstrates moderate to high reliability and validity, and high internal consistency (Cronbach’s alpha for all subscales ≥0.92) (Lovibon & Lovibond, 1995).
Table 4.1 DASS21 scores and corresponding descriptors

<table>
<thead>
<tr>
<th></th>
<th>Normal</th>
<th>Mild</th>
<th>Moderate</th>
<th>Severe</th>
<th>Extremely Severe</th>
</tr>
</thead>
<tbody>
<tr>
<td>Depression</td>
<td>0-9</td>
<td>10-13</td>
<td>14-20</td>
<td>21-27</td>
<td>28+</td>
</tr>
<tr>
<td>Anxiety</td>
<td>0-7</td>
<td>8-9</td>
<td>10-14</td>
<td>15-19</td>
<td>26-33</td>
</tr>
<tr>
<td>Stress</td>
<td>0-14</td>
<td>15-18</td>
<td>19-25</td>
<td>26-33</td>
<td>34+</td>
</tr>
</tbody>
</table>

Clinician Questionnaires

The clinician questionnaire included the following purpose-designed measures, to gain quantitative and qualitative feedback on the booklet:

General professional demographics: a purpose-designed questionnaire was used to elicit participants’ demographic and professional characteristics including their clinical field/specialty, and years of experience with GA disorders.

Booklet Feedback Questionnaire: this purpose-designed questionnaire was used to assess the clinician’s views on the booklet’s usefulness, perceived benefit, and acceptability. The questionnaire was similar to the patient group questionnaire as described above, with wording changes as appropriate to the clinician group. The questionnaire also consisted of 13 short-answer questions about the clinician’s initial impression, content, layout, and usability of the booklet, and 12 statements with five response options ranging from “Strongly Disagree” to “Strongly Agree”.
4.4.5 Qualitative Measures

*Semi-structured Interviews*

Semi-structured interviews with the patient group were conducted by the student researcher. Interviews were conducted via phone with participants who returned a consent and contact form by mail and who indicated a willingness to participate in an interview. Alternatively, if participants indicated that they would be willing to be interviewed when they were attending the clinic for an appointment, a researcher conducted a face-to-face interview after their medical consultation.

The interview questions were designed to gather feedback about the booklet in line with the aims of the research – to assess the booklet’s clarity, usefulness, and acceptability. Participants were asked open-ended questions about their overall likes and dislikes of the booklet, and then more specifically about each of the main three sections of the booklet (described in Chapter 3.2). Participants were also asked if there were information or other elements in the booklet that they would like to see included or omitted. The responses were noted verbatim by the researcher and transcribed to a spreadsheet under each question. The Interview Script is available in Appendix G.

4.4.6 Data Analysis

*Quantitative data*

Quantitative data from the standardised and purpose-designed scales were used to obtain feedback about participant perception of the booklet, and demographic and clinical characteristics. The data was entered into SPSS (IBM SPSS Statistics Premium, Version 19.0) and analysed using descriptive statistics.
Qualitative analysis

Thematic analysis was used to analyse the qualitative data gathered in this study. Thematic analysis is used to identify, analyse, and report identified patterns within the data (Braun & Clarke, 2006), aiming to map out the range of concepts or summarise unstructured data in thematic categories. Thematic analysis was used in a deductive way in this study, in that coding and theme development were directed by the existing concepts and categories relating to the feedback about the booklet.

Combining thematic analysis with content analysis is a useful strategy (Leech & Onwuegbuzie, 2008). Content analysis captures the frequency of codes in order to determine the concepts most raised in the data. Rosenthal and Rosnow (1991) defined content analysis as a method of categorising information through the use of frequency of occurrences. In the health sciences, content analysis is accepted and widely used as a standard qualitative research method (Hsieh & Shannon, 2005). With its focus on communication (Downe-Wamboldt, 1992), content analysis is suitable for patient health education and clinical practice, and therefore most relevant to the current study.

The data collected in the current study were categorised using a priori coding (deductive approach), however data were also given a new category based on latent or emergent meanings if necessary. Responses from each question of the interviews and study questionnaires (short answer/open ended questions) were entered into a spreadsheet with the frequencies of responses recorded. If responses were better suited to a broader category that emerged during the analysis, they were allocated to those new categories.
4.5 Results

4.5.1 Patient Group

Sample characteristics

Approximately 80 potential participants were invited to take part in the study. Twenty-six questionnaires were returned to the research team, representing a 31% response rate. Six questionnaires were excluded due to incompleteness of responses or ineligibility, leaving twenty questionnaires. A relatively low number of participants was anticipated given the scarcity of GA disorders and that patients often attend specialist appointments only annually, therefore the opportunities to access patients was restricted. In addition, this was an opt in study requiring mail back of the completed questionnaire. This may have been a barrier to a greater response rate even though potential participants indicated initial interest.

Table 4.2 shows the demographic and clinical characteristics of the patient group. The sample consisted of 50% female and 50% male participants. The average age of participants was 49 years (SD 14.3), ranging between 22 and 73 years. The average number of children of participants was two (SD 1.2), ranging between zero to four. The majority were married, and the highest level of education was postgraduate degree. The main country of birth was Australia, and all participants spoke English at home.
Table 4.2 Demographic characteristics of the patient group

<table>
<thead>
<tr>
<th>Variable</th>
<th>n</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Relationship status</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Never married</td>
<td>2</td>
<td>10</td>
</tr>
<tr>
<td>Married/De facto</td>
<td>16</td>
<td>80</td>
</tr>
<tr>
<td>Widowed</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Divorced/Separated</td>
<td>2</td>
<td>10</td>
</tr>
<tr>
<td><strong>Highest Education Level</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Year 12/HSC/Leaving certificate or below</td>
<td>4</td>
<td>20</td>
</tr>
<tr>
<td>TAFE certificate/diploma</td>
<td>4</td>
<td>20</td>
</tr>
<tr>
<td>University degree</td>
<td>6</td>
<td>30</td>
</tr>
<tr>
<td>Higher degree (postgraduate)</td>
<td>6</td>
<td>30</td>
</tr>
<tr>
<td><strong>Country of Birth</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Australia</td>
<td>13</td>
<td>65</td>
</tr>
<tr>
<td>UK/Europe</td>
<td>5</td>
<td>25</td>
</tr>
<tr>
<td>USA</td>
<td>1</td>
<td>5</td>
</tr>
<tr>
<td>NZ</td>
<td>1</td>
<td>5</td>
</tr>
<tr>
<td><strong>Language spoken at home</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>English</td>
<td>20</td>
<td>100</td>
</tr>
</tbody>
</table>

Clinical Characteristics of the Sample

Table 4.3 shows the clinical characteristics of the sample. The average time since diagnosis was 11 years (SD 10.4), ranging between 0-27 years. Thirteen (65%)
participants had undergone surgical treatment, and some had undergone multiple surgeries.

Table 4.3 Clinical characteristics of the patient group

<table>
<thead>
<tr>
<th>Surgery</th>
<th>n</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>None</td>
<td>7</td>
<td>35</td>
</tr>
<tr>
<td>Stent graft</td>
<td>2</td>
<td>10</td>
</tr>
<tr>
<td>Valve replacement</td>
<td>4</td>
<td>20</td>
</tr>
<tr>
<td>Other (non-aortic)</td>
<td>7</td>
<td>35</td>
</tr>
<tr>
<td>If had surgery, preventative</td>
<td>6</td>
<td>30</td>
</tr>
<tr>
<td>If had surgery, emergency</td>
<td>2</td>
<td>10</td>
</tr>
</tbody>
</table>

Information Preferences

Most participants preferred to receive ‘as many details as possible’ about their condition, with an average rating of 9 out of 10 (range 6-10, SD=1.3).

Table 4.4 shows information sources accessed by participants. All participants sought information about their condition from at least one health or medical related Internet site, and one quarter (25%) specifically indicated accessing information through their medical specialist in person (e.g. cardiologist, genetic counsellor). Information sheets were accessed for associated conditions, for example hypertension management.
Table 4.4 Information sources accessed

<table>
<thead>
<tr>
<th>Information source accessed</th>
<th>n</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Health or medical related website</td>
<td>20</td>
<td>100</td>
</tr>
<tr>
<td>Book</td>
<td>2</td>
<td>10</td>
</tr>
<tr>
<td>Information sheet</td>
<td>4</td>
<td>20</td>
</tr>
<tr>
<td>Medical professional</td>
<td>5</td>
<td>25</td>
</tr>
</tbody>
</table>

*Psychological Well-being*

Table 4.5 shows the psychological well-being profile of the sample. On the Depression, Anxiety and Stress Scale (DASS21; Lovibond & Lovibond, 1995), overall patient sample scores fell in the Normal range on the depression subscale, anxiety subscale, and stress subscales indicating that there were no clinical concerns relating to depression or anxiety amongst the participants.

Table 4.5 Psychological well-being profile

<table>
<thead>
<tr>
<th>DASS21 (possible range)</th>
<th>Mean</th>
<th>SD</th>
<th>Qualitative reference (range)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Depression (0-28+)</td>
<td>1</td>
<td>1</td>
<td>Normal range (0-9)</td>
</tr>
<tr>
<td>Anxiety (0-20+)</td>
<td>2</td>
<td>3</td>
<td>Normal range (0-7)</td>
</tr>
<tr>
<td>Stress (0-34+)</td>
<td>2</td>
<td>2</td>
<td>Normal range (0-14)</td>
</tr>
</tbody>
</table>
Quantitative Feedback about the Booklet

Overall, the booklet was very positively endorsed by participants for content, format and acceptability. The most endorsed items were: the booklet being easy to understand (n=20/20), and the overall helpfulness of the booklet (n=18/20). The least endorsed item was whether the participant would bring the booklet to future consultations with the clinician (n=10/20 were unsure).

Content of the booklet

Table 4.6 shows the frequency of responses to each item on the Feedback Scale relating to the content of the booklet. The majority of participants rated each item relating to the content of the booklet positively (‘Strongly Agree’ or ‘Agree’). Participants highly endorsed that the booklet was easy to understand (n=20/20), that the booklet was helpful overall (n=18/20), and that the booklet answered questions about how the GA disorder would affect them (n=19/20). The least endorsed item was the usefulness of the section on coping with the emotional impact (n=13/20 agreed or strongly agreed).
Table 4.6 Feedback about the content of the booklet (frequency of responses)

<table>
<thead>
<tr>
<th>Statement</th>
<th>Strongly Disagree/Disagree n (%)</th>
<th>Unsure n (%)</th>
<th>Agree/Strongly Agree n (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>The booklet was easy to understand</td>
<td>0</td>
<td>0</td>
<td>20 (100)</td>
</tr>
<tr>
<td>The booklet was helpful to me overall</td>
<td>1 (5)</td>
<td>1 (5)</td>
<td>18 (90)</td>
</tr>
<tr>
<td>The booklet answered questions about how my condition may affect me</td>
<td>0</td>
<td>1 (5)</td>
<td>19 (95)</td>
</tr>
<tr>
<td>After reading the booklet, I know more about what to expect with treatment</td>
<td>0</td>
<td>5 (25)</td>
<td>15 (75)</td>
</tr>
<tr>
<td>The booklet provided information on how to best care for myself</td>
<td>0</td>
<td>3 (15)</td>
<td>17 (85)</td>
</tr>
<tr>
<td>The section on Coping with the emotional ups and downs (p.17) was useful</td>
<td>5 (25)</td>
<td>2 (10)</td>
<td>13 (65)</td>
</tr>
</tbody>
</table>

Format of the booklet

The majority of participants endorsed the format of the booklet, strongly agreeing that the booklet was easy to read (n=20/20), with the majority liking the design/look of the booklet (n=19/20). The majority also indicated that the booklet was the right length (n=16/20). Table 4.7 shows the frequency of responses to each item on the Feedback Scale relating to the format of the booklet.
Table 4.7 Feedback about the format of the booklet (frequency of responses)

<table>
<thead>
<tr>
<th>Statement</th>
<th>Strongly Disagree/Disagree n (%)</th>
<th>Unsure n (%)</th>
<th>Agree/Strongly Agree n (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>The booklet was easy to read</td>
<td>0</td>
<td>0</td>
<td>20 (100)</td>
</tr>
<tr>
<td>I liked the look of the booklet</td>
<td>0</td>
<td>1 (5)</td>
<td>19 (95)</td>
</tr>
<tr>
<td>The booklet was the right length</td>
<td>2 (10)</td>
<td>2 (10)</td>
<td>16 (80)</td>
</tr>
</tbody>
</table>

Acceptability of the booklet

Most participants (n=18/20) indicated that they did not feel anxious while reading the booklet. While half of the participants were not sure if they would bring the booklet to future appointments, a quarter of the sample indicated that they would. Almost all participants would recommend the booklet to other people with a GA disorder (n=19/20). Table 4.8 shows the frequency of responses to each item on the Feedback Scale relating to the acceptability of the booklet.
Table 4.8 Feedback about the acceptability of the booklet (frequency of responses)

<table>
<thead>
<tr>
<th>Statement</th>
<th>Strongly Disagree/Disagree</th>
<th>Unsure</th>
<th>Agree/Strongly Agree</th>
</tr>
</thead>
<tbody>
<tr>
<td>I felt anxious while reading the booklet</td>
<td>18 (90)</td>
<td>1 (5)</td>
<td>1 (5)</td>
</tr>
<tr>
<td>I would bring and refer to this booklet in future consultations with my physician</td>
<td>5 (25)</td>
<td>10 (50)</td>
<td>5 (25)</td>
</tr>
<tr>
<td>I would recommend this booklet to other people with a genetic aortic disorder</td>
<td>0</td>
<td>1 (5)</td>
<td>19 (95)</td>
</tr>
</tbody>
</table>

Topics of Most Importance

Participants were given the opportunity to give free text responses to the survey statement: “The information of most importance to me were: (Please indicate why)”. The following categories demonstrate the pattern of responses.

Treatment topics: Participants strongly endorsed the importance of the information in the booklet about medical management of GA disorders and the associated treatment, including graphics illustrating surgical repair: “[The important booklet topic was] treatment and what to expect because you tend not to want to ask the specialists too many questions”, “Aortic aneurysms can be repaired. It is comforting information”, “Illustration of aortic root repair and how it was stitched.”
In particular, the information relating to emergency medical management was considered very important: “What to do in an emergency, everybody should be given a medical management plan”.

A variety of lifestyle and practical topics were also considered very important, including information about lifestyle changes: “It was useful to know what I myself could do to prevent problems”, “Lifestyle and exercise, as exercise is particularly important to me”, and commonly asked questions: “Myths and misconceptions also very useful”

Participants supported the sections on genetic related information as clear, necessary, and supportive: “Testing of other family members [is] now clear”, “Since GA is inherited, it would be wise to advise for family members to get tested. This advice saved my father’s life”, “What a brilliant resource! Fantastic, I wish it had been there 20 years ago when I was a teenager being told that I needed open heart surgery. My son has inherited Marfans from me, and when the time comes that he has to face up to his Marfans in terms of surgery, I can honestly say that this book would form an integral part of support him (and the whole family) through the process”.

Reported topics of least importance were those not medically relevant to the participant. For example, a participant who did not undergo surgery wrote: “Aortic dissection was not involved in my care”.
Participants found the emotions and coping section less useful if they thought that coping should not be an issue: “The information regarding the emotional impact on people as it is not something I dwell on. I always think of people in worse conditions”, “Emotional ups and downs, as I’m fairly resilient and practical”, “Coping with emotional ups & downs. Why? What happens, happens!!”

Feedback from participants about their perceived strengths of the booklet included positive comments about the format and layout: “Well set out easy to read and understand”, “It's an easy book to read for anyone - easy to understand in layman’s terms”, its usefulness for specific patients: “I felt it could all be useful to a newly diagnosed patient”, and for knowledge gain: “An excellent initiative that should be pursued for patients - one of the greatest sources of anxiety is fear of the unknown. This booklet goes some way to addressing that issue”.

Suggestions for additional topics related to including more detail about surgical treatment and post-operative issues, and greater detail about genetic issues in the booklet.

Suggestions for improvement included, to include a list of acronyms, and to provide access to an online version of the booklet with links to scientific journal articles.
4.5.1 Participant Interviews

The researcher conducted seven phone or face-to-face interviews with additional participants for more in-depth feedback about the usefulness, format, and acceptability of the booklet. The qualitative feedback (from the interviews) largely reflected the quantitative feedback obtained through the questionnaires. However, the interviews gave opportunities to clarify patterns of responses in the questionnaires, for example, the divergent responses to the topic of emotional coping. Direct quotes from participants are italicised and enclosed in quotation marks.

Booklet first impressions

First impressions about the booklet were unanimously positive. Participants commented about the informative content, the good flow of the information, and clean, clear layout: “[The booklet is] very well set out, explained lots of things, quite good information”, “Good, compact, informative, not a catalogue of info”. The convenience of the information was appreciated, in that the information was effortlessly available: “All the info is in an accessible spot. I tried to look up info before [online], it was difficult”.

Information dissemination

The participants appreciated that the booklet was written in lay language: “It explained information without complicated information”, “Good booklet, outlines disorders [in] simple description”. All participants agreed that the information was easy to comprehend: “Very good breadth of information, informative to the average person”.
without medical knowledge”, with the medical terminology easy to understand in comparison to other sources: “On the Internet, [it is] too hard to understand the words”.

The feedback indicated that the content was set out in a progressive way with information encouraging evolving knowledge: “Flowed really well: content, intro, related to the story, steps through each section very well”, “[The booklet] develops as it goes, with succinct process to explain conditions and solutions”. The feedback also indicated that the booklet provided a bridge between scientific information that patients have researched and the recommended treatment plan: “[The booklet] provides a breakdown of barrier between researchers and what doctors tell you”.

One participant commented that the brief but succinct descriptions of the GA disorders could be used to relay relevant information to other people, such as family members or friends: “Short description [of the conditions] was useful, good to use to explain to someone else”.

Usefulness

Participants indicated that the booklet would be useful for a person recently diagnosed with a GA disorder. It was suggested that the booklet gave an explanation of the conditions and other associated issues that form a base on which the individual can review and build up more knowledge. “Yes, I get more information from the booklet, what can happen, who to see, helps the person, it’s important”. “[The booklet] great
for outside the appointment. It gives basic understanding, you can then ask questions to the doctor. Can build up and research more info”.

Further, one participant commented that the booklet would aid in the understanding of topics as she did not always ask her clinician about some issues and: “Yes, it explains things, sometimes I don’t ask doctor about things”.

Timing

In terms of when it would be appropriate for patients to be given the booklet, participants supported the use of the booklet in the first consultation or early on in the illness, as a supportive resource for an individual receiving a diagnosis: “[The booklet should be given at] the first appointment, as it fills in missing information I didn’t ask the doctor” or “Within six weeks after, in the first checkup; [since if given] too early not good, as you can’t absorb”. In addition, participants suggested that the booklet was useful as a reference at a later time outside of consultations: “Information in appointments are overwhelming, you can look at info [in the booklet] later with a clear head”.

Support from the treatment team

Having a member of the treatment team go through parts of the booklet with a patient was considered a useful complementary use of the booklet: “Have conversation about condition then go away with booklet and option to call or follow up if have questions”, although some did not think that it would a necessary strategy as the booklet was deemed a standalone resource which was easy to read and understand: “[the booklet] is self-
explanatory””. However, one participant acknowledged that the booklet was not intended to replace direct medical advice: “[The booklet provides] basic outline, for any more information you should ask your doctor”.

Design and layout of the booklet

The layout of the booklet in terms of its size, length, flow, colour and look were positively reviewed: “Size and colour of the booklet is good, it makes you want to read it - colourful, it stands out”, “[The booklet is] not too big, it is to the point, you don’t need too much information, otherwise it gets confusing”.

Participants commented that the graphics were relevant and supported the text: “Good, ideal images, it is nice to see what an aneurysm is, it clarifies text” or “Very appropriate pictures that explain text”.

One participant commented that the tone of the booklet was appropriate and enhanced his sense of hope: “[The booklet was] optimistic, even though I have Marfans”.

Specific sections of the booklet

Participants were asked to comment about specific sections of the booklet.

The ‘Patient’s story’ was well-received, with most participants indicating that they related to parts of the story: “I related to the story – in terms of age, I recall Prof saying about weight training. Not so much the attitude, but to elements of emotional journey and
having positive outlook”, “[The story was] good, it gives understanding of other’s’ journey and positive outcome, it helps with basic understanding”.

The feedback about the medical information section was that it gave a good description of GA disorders, related issues, and treatment options: “Yes, different conditions were covered, good to know, statistics is good” or “Good, it gives description, pictures are helpful, explains dissection well”.

The person-oriented section about lifestyle, frequently asked questions, and coping were appreciated by those interviewed. Comments included: “Lifestyle changes are important and relevant”. Participants particularly valued the ‘Myths and Misconceptions’ section: “especially Myths and Misconceptions about life expectancy”. One participant commented that these topics aided in her self-management: “Helps me to help myself”.

Similarly to the quantitative responses, participants offered a variety of opinions about the ‘Coping with Emotions’ section: “Coping section good”, “Not sure”, “Coping – identify with it and might use it”. Upon further enquiry, it appeared that participants liked the topic if they were oriented towards exploring their emotional needs: “Coping with the diagnosis. Normalising”, or disliked it if they tended towards logical problem-solving and coping styles: “Emotions section – can see why it is there, but not for everyone. Put it after questions”.
Respondents liked the practicality of the section to document their appointments, test results, and pages with example questions to ask the clinician and links to resources: “Helpful to remember appointments”, “Yes, in case I forget, and to remember what was said in the appointment”, “Good to have it to choose to use it or not. Would use it as a record”.

Participants were asked about their most liked topics in the booklet. Participants nominated a variety of topics, such as surgery: “Parts about surgery, risks, types of surgery”, and genetic information: “Information about genetic info”. The feedback also indicated that the ‘Questions Checklist’ was considered useful as a reminder or prompt for questions to ask or topics to discuss in consultations: “I have asked some of these questions before, but saw some in the booklet that I would like to ask”, “Questions to ask, use as prompts, reminder”. No participants indicated that they disliked any part of the booklet with most responses as “nothing” or “nil”.

As GA disorders have a familial component, often individuals need to consider the impact of the diagnosis on family members. Half of the participants indicated that their family members had read the booklet, and that they had found it useful: “Yes, it is information-giving, it explains things, and is good to have it on hand [for family members]”. The other half of the respondents indicated that they would give other family members the booklet to read: “I related to “Don’t want to worry my family” [in the text of the booklet in coping section]”, “Yes. [The booklet]would be good for the adjustment period, it would be a starting point, to start the conversation [with others]”.
Participants were given the opportunity to make any other comments about the booklet. Comments included that the booklet was considered valuable as it was endorsed by the clinician: “Fantastic. Would have made it easier at first [to have the booklet], because there’s a long time between appointments. If a doctor hands [the booklet] to you, keep it”. The booklet also helped to facilitate a sense of control over the condition and treatment: “Can manage condition rather than endure it, not as a passenger in a medical journey”, “[Information in the booklet] gives options, can control aspects of your life”, and that it was a helpful resource: “Very informative and helpful booklet”.

4.5.3 Clinician Group

Sample Characteristics

Table 4.9 shows the demographic and professional characteristics of the clinician group. The clinician sample (n=9) included two female, and seven male respondents, with all participants practising as cardiologists or surgeons. The average number of years of clinical experience was 16 years, ranging from 10 to 30 years. The participating clinicians mostly worked in both public and private settings (concurrently), and usual patient diagnosis was Bicuspid aortic valve disorder, followed by Marfans Disease. Only one respondent said that they used patient information materials in their consultation with patients.
Table 4.9 Demographic and professional characteristics of the clinician group

<table>
<thead>
<tr>
<th>Variable</th>
<th>n</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Gender</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Male</td>
<td>7</td>
<td>78</td>
</tr>
<tr>
<td>Female</td>
<td>2</td>
<td>22</td>
</tr>
<tr>
<td><strong>Clinical Field</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Cardiologist</td>
<td>8</td>
<td>89</td>
</tr>
<tr>
<td>Surgeon</td>
<td>1</td>
<td>11</td>
</tr>
<tr>
<td><strong>Service Type (can be multiple types)</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Public</td>
<td>7</td>
<td>78</td>
</tr>
<tr>
<td>Private</td>
<td>7</td>
<td>78</td>
</tr>
<tr>
<td>Community</td>
<td>2</td>
<td>22</td>
</tr>
<tr>
<td><strong>Most common GA Disorders in patient group (can be multiple)</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Bicuspid Aortic Valve</td>
<td>7</td>
<td>78</td>
</tr>
<tr>
<td>Marfans Disease</td>
<td>4</td>
<td>44</td>
</tr>
<tr>
<td>TAAD</td>
<td>1</td>
<td>11</td>
</tr>
<tr>
<td>GA Disorder (general)</td>
<td>1</td>
<td>11</td>
</tr>
<tr>
<td>Other</td>
<td>1</td>
<td>11</td>
</tr>
<tr>
<td><strong>Age range of patients</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>All adolescents upwards</td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Mean</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Range</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>SD</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>No. of years of clinical experience</td>
<td>14.7</td>
<td>10-30</td>
</tr>
<tr>
<td>Current use of patient psycho-education booklets or other educational material</td>
<td>Yes (n=1)</td>
<td>No (n=8)</td>
</tr>
<tr>
<td>1 (Pamphlet, type not specified)</td>
<td>8</td>
<td></td>
</tr>
</tbody>
</table>
When asked why written material was not used. The majority of participating clinicians cited the lack of availability of, or accessibility to, evidence-based materials: "Not available to me. Send me more booklets!", "Not available. Unaware of such material", and "Do not have access to a booklet".

Quantitative feedback about the booklet

Overall, the majority of the clinicians strongly agreed that the content of the booklet was useful and appropriate, the booklet was easy to read, understand, and provided adequate information about treatment. The majority liked the look of the booklet, and indicated that the booklet was the right length.

Acceptability of the booklet

The majority of the clinician group agreed that the booklet was appropriate for the target audience, and would give the booklet to patients with a GA Disorder. Table 4.10 shows the frequency of responses to each item on the Feedback Scale relating to the acceptability of the booklet.
Table 4.10 Feedback about the acceptability of the booklet (frequency of responses)

<table>
<thead>
<tr>
<th>Statement</th>
<th>Strongly Disagree/ Disagree n (%)</th>
<th>Unsure n (%)</th>
<th>Agree/ Strongly Agree n (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>The information in the booklet was appropriate for the target audience</td>
<td>1 (11)</td>
<td>0</td>
<td>8 (89)</td>
</tr>
<tr>
<td>The sections that allow space to write notes would be useful in consultations, e.g. appointments, test results, questions, notes</td>
<td>2 (22)</td>
<td>1 (11)</td>
<td>6 (67)</td>
</tr>
<tr>
<td>The Questions to Ask Your Physician checklist (p.20) would be useful to refer to in consultations</td>
<td>0</td>
<td>1 (11)</td>
<td>8 (89)</td>
</tr>
<tr>
<td>I would give this booklet to patients with a genetic aortic disorder</td>
<td>1 (11)</td>
<td>0</td>
<td>8 (89)</td>
</tr>
</tbody>
</table>

Content of the booklet

Table 4.11 shows the frequency of responses to each item on the Feedback Scale relating to the content of the booklet. The booklet content was considered to be useful and gave adequate information for the target audience (n=8/9).
Table 4.11 Feedback about the content of the booklet (frequency of responses)

<table>
<thead>
<tr>
<th>Statement</th>
<th>Strongly Disagree/Disagree n (%)</th>
<th>Unsure n (%)</th>
<th>Agree/Strongly Agree n (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>The booklet was easy to understand</td>
<td>1 (11)</td>
<td>0</td>
<td>8 (89)</td>
</tr>
<tr>
<td>The information in the booklet would answer patients’ questions about how their condition may affect them</td>
<td>1 (11)</td>
<td>0</td>
<td>8 (89)</td>
</tr>
<tr>
<td>The booklet provided adequate information about treatment</td>
<td>1 (11)</td>
<td>0</td>
<td>8 (89)</td>
</tr>
<tr>
<td>The information about making lifestyle changes would be useful for patients</td>
<td>0</td>
<td>0</td>
<td>9 (100)</td>
</tr>
<tr>
<td>The sections on coping, myths, and FAQs would be useful for patients</td>
<td>0</td>
<td>1 (11)</td>
<td>8 (89)</td>
</tr>
</tbody>
</table>

**Format of the booklet**

The majority of clinicians agreed/strongly agreed that the booklet was easy to read (n=8/9) and all agreed/strongly agreed that the booklet was the right length (n=9/9). Table 4.12 shows the frequency of responses to each item on the Feedback Scale relating to the format of the booklet.
Table 4.12 Feedback about the format of the booklet (frequency of responses)

<table>
<thead>
<tr>
<th>Statement</th>
<th>Strongly Disagree/Disagree n (%)</th>
<th>Unsure n (%)</th>
<th>Agree/Strongly Agree n (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>The booklet was easy to read</td>
<td>1 (11)</td>
<td>0</td>
<td>8 (89)</td>
</tr>
<tr>
<td>The booklet was the right length</td>
<td>0</td>
<td>0</td>
<td>9 (100)</td>
</tr>
<tr>
<td>I liked the look of the booklet</td>
<td>1 (11)</td>
<td>2 (22)</td>
<td>6 (66)</td>
</tr>
</tbody>
</table>

Qualitative feedback about the booklet

Qualitative feedback from the clinician group about the booklet related to their initial impressions, use, and content of the booklet.

First Impressions: Clinicians’ first impressions of the booklet were positive, referring to the quality, and overall usefulness of the booklet: "An excellent comprehensive tool", "Very good, nice and clear", and "Great resource".

Design and Layout: The design and layout of the booklet was regarded positively:
"Design is good", "Nice clear design. Good use of colour to highlight important graphics".
Usefulness: Clinicians supported the use of the booklet as an psycho-education booklet for a person who has recently (within 6 months) received a diagnosis of a GA disorder due to the appropriateness of the included topics as a starting point of educating the patient: “[The booklet] provides clear information in simple terms”, "It is informative and can aid to understanding”, and "Good source of initial information”.

Clinicians also approved of the booklet for use as an information tool in consultations, given time constraints and usefulness of the booklet as a reference source away from the consultation: "[The booklet] provides more information than time would otherwise permit”, "It fills in the gaps as consultations usually focus on one or two areas. It will also reinforce what has been discussed”, and "It takes the pressure off as they can take it and read at leisure”.

The majority of clinicians (n=8 out of 9) suggested that the physician or nurse from the treating team should go through some of the booklet pages with the patient before it was taken home.

The majority of clinicians reported that the booklet would be useful for most of their patients: "All patients with aortic disorder","Young patients, patients with a reasonable level of education”, “20-60 year old”, except those with low level English literacy or comprehension.
Weaknesses: Criticisms of the booklet were also noted by one clinician, and included that the booklet: "Starts with a disaster story. Typical hospital perspective" and was "Too high pitched. Not reassuring".

Additional Topics: Suggestions for additional topics to be included in the booklet included: information about the safety of sex; criteria for surgery; further information sources; information about carrying a medical tag.

There were no suggestions for the removal of any topics.
CHAPTER 5: DISCUSSION

5.1 Research Aims

This study aimed to develop and evaluate a psycho-education booklet designed specifically for patients recently diagnosed with a GA disorder. The main aims were: to assess the booklet’s clarity, usefulness, and acceptability by the patient population and by clinicians specialising in GA disorders.

5.2 Discussion of the main findings

The overall findings of the evaluation indicated that the information booklet was very well-received by the patient group. The feedback from the patient group was very positive about the concept of the booklet, which was considered practical and informative. Overall, patients found the booklet easily understandable, and of the right length, depth and breadth. The look, presentation, and size of the booklet was also considered appropriate and enhanced the booklet’s usability.

Overall, the surveyed clinicians endorsed the booklet as an useful information resource about GA disorders and associated topics. The clinician group supported giving the booklet to patients at the time of diagnosis, and its ongoing use in future consultations. The majority of clinicians commended the booklet content as suitably pitched for the intended audience, and the format as appropriate in length, booklet size, with clear layout and graphics.
Supporting patients to be active participants in their treatment

Studies have shown that patients often passively engage in their own care (Nolte & McKee, 2008), and that appropriately designed resources are needed to facilitate patients to have a more active involvement and effective communication with health professionals (Dowie, 2002). As Hagerty et al. (2005) reported, patients commonly have a limited understanding of their diagnosis, prognosis and treatment plans. The study booklet is a resource that was designed to provide easily understood medical information about complex conditions and treatment. Clinicians commented that the range of topics covered in the booklet would aid patients to understand how the GA disorder may affect them, and help them in making treatment decisions and adhering to treatment plans. The patients surveyed and interviewed in this study confirmed that the content was easy to read and understand. It was shown that the booklet described in this thesis has the potential to assist in educating patients and their support persons about GA disorders, treatment, and lifestyle changes. In addition, the written information could complement and further reinforce the verbal information given in medical consultations.

Participants in the patient group that had been newly diagnosed with GA disorder rated the information in the booklet as an excellent overview and starting point from which to gather more information via their medical team or other sources. Whilst there is a paucity of research specifically about information dissemination for patients with GA disorders, research has found that recall and knowledge are enhanced by written information in individuals with a cancer diagnosis, and that practical information presented in booklets are valued (McPherson et al., 2001). In terms of practical content, the overall patient
group rated emergency planning as an important and valuable section as it was a major and continual concern for patients. In addition, having easily accessible information that could be personalised to the individual’s needs, and instructions for support or medical persons was considered an easy and practical solution.

In addition, the list of “Questions to Ask Your Physician” (the booklet’s ‘Question Prompt List’, QPL) was regarded by the patient group as a practical communication aid that would alert or remind them of issues to discuss with their clinician. Patients who participated in the interviews commented that the QPL would be particularly helpful as a reminder of questions to ask and as a checklist of forgotten or further questions to ask their clinician. In addition, the clinician group commented that the QPL could prompt patients at a later time when the initial impact of the diagnosis had lessened. QPLs can increase the communication between patient and clinician, and is most effectively done when the use of the QPL is promoted by the clinician and questions are specific to the medical issue (Sansoni, Grootemaat, & Duncan, 2015). Sansoni, Grootemaat, and Duncan’s (2015) review of the use and effectiveness of QPLs found that in general, patients asked more questions and that clinicians gave more information when a QPL was provided and used. The review recommended that QPLs should include questions about treatment, test options, the risks and benefits, and the likelihood of those occurring. The use of QPLs has been found to increase the questions asked by patients in areas such as prognosis (Brown, Butow, Dunn, & Tattersall, 2001), diagnosis (Bruera, Sweeney, Willey, Palmer, Tolley, Rosales, & Ripamonti, 2003), and treatment issues (Smets, van Heijl, van Wijngaarden, Henselmans, & van Berge Henegouwen, 2012). In terms of
when to provide a QPL, Sansoni et al. (2015) suggested that a QPL be best provided at diagnosis or early stage of a phase of treatment. This is supported by the current study’s booklet design for patients newly diagnosed with a GA disorder.

The patient group in the current study self-rated highly on the information preference scale, meaning that they identified themselves as ‘active seekers’ of information about their condition, treatment and associated issues. No other studies are available for comparison that have investigated the information preferences of patients specifically with a GA disorder. However, it has been shown in various studies of individuals diagnosed with a chronic illness that patients tend to rate highly on information-preference, meaning that they prefer to have as much information as possible in order to participate in decision-making about treatment (e.g. James et al., 1999; Cartwright & Windsor, 1993).

Individuals with high information-preference levels prefer to have detailed information about the condition, treatment options, impact on lifestyle, and participate in shared decision-making with the treatment team. Research on the factors that influence patient information preferences and involvement in treatment decisions have yielded variable findings. These include factors such as socio-demographic variables, experience with health care, health status, and the type of decision (Say, Murtagh, & Thomson, 2006; Deber, Kraetschmer, & Irvine, 1996; Mansell, Poses, Kazis, & Duefield, 2000). Arora and McHorney’s (2000) observational study of patients with chronic disease including myocardial infarction, congestive heart failure, and depression, showed that patients who
preferred more information and a greater role in decision-making, were more likely to use active (versus passive) coping strategies. It may be the case that the participants in the current study tended to be individuals with active coping strategies. Being a highly educated sample may have also been an influence on the greater information-seeking preference of the patient group and self-selection into the study. There is a need to explore the information preferences of a larger and more varied sample of patients diagnosed with a GA disorder. Tailoring information resources based on the patient’s preference for the amount of detail they wish to receive, for example adding optional information to the booklet, may also facilitate adherence to treatment and self-management (Shepherd, Barratt, Trevena, McGeechan, Carey et al., 2011).

Although the booklet was designed for individuals with a recent diagnosis of a GA disorder, it was anticipated that feedback from a broad range of GA patients, varying in time since diagnosis, would yield valuable information. Participants more experienced in their medical journey (i.e. in follow up) indeed gave important opinions from a retrospective standpoint and strongly endorsed the use of the booklet at the earliest time from diagnosis. The feedback from patients with a shorter time since diagnosis also supported for the booklet to be available at the first (or early) appointments following the diagnosis. All patients also indicated that the study booklet would be useful to have available, and to refer back to, throughout the treatment. It was suggested that the booklet would help to bridge the gap between what was said, heard, and remembered in the appointment and what was forgotten to ask. Furthermore, giving the booklet to family members or friends was suggested as an additional use for the booklet. While the current
study did not assess longer-term outcomes, other studies have shown longer-term benefits with the provision of written health information such as improved coping, increased self-management, and a greater sense of control (Mills & Sullivan, 1999; Treacy and Mayer, 2000; Chelf et al., 2001).

Similarly, clinicians suggested that the most appropriate time to give patients the booklet would be at the time of diagnosis or at the early stages of treatment, as the booklet gave a good overview of GA conditions, treatment, impact, and associated lifestyle issues. The booklet may assist patients who may not have fully understood the information given at the time of the consultation or have questions they had forgotten to ask. The booklet could then serve as an ongoing information support resource for the patient, clinician, and the patient’s support persons.

The majority of the surveyed clinicians indicated that they would give the booklet to patients as well as encourage patients to refer to it between consultations. However, patients gave mixed responses on whether it would be useful if a member of the medical team highlighted relevant information in the booklet during a consultation. It was suggested by patients that this may be useful but not a necessary step. This reflects the varied findings in the literature about the benefits of verbal discussion by a health professional of written material. No knowledge gain was found with written information together with verbal discussion (Lowe, Sharma, & Leathley, 2007), however, patient satisfaction was reported to increase with written information complementing verbal information (Dunnill & Pounder, 2004). Therefore, the findings of the current and
previous studies suggest that the option of having a clinician explain parts of the booklet may be best determined by the needs and preferences of the patient.

Both patient and clinician participants also varied in whether they would make use of the booklet in subsequent medical consultations, although most interviewed patients reported that it was likely that they would refer to the booklet on an ongoing basis. Therefore, it seems likely that the continued use of the booklet would also be based on the patient’s personal preference. Having the GA disorders booklet continuously available is an important option. It has been shown that printed materials supplemented by verbal information given by health professionals is a frequently used and preferred method of knowledge gain by patients (Rutten et al., 2005).

5.2.1 Usefulness and acceptability

All but one clinician surveyed reported that they had never provided printed information about GA disorders to their patients. The most common reason given was the perceived lack of appropriate and specific patient information resources available, hence the study’s booklet would be a needed and welcomed resource in clinical practice. This supported the findings of the needs assessment, namely that the there was a lack of appropriate and easily accessible psycho-education resources for patients with a GA disorder and that patients would like specific information on GA disorders, treatment, and lifestyle impact (Connors et al., 2015).
The Development and Evaluation of a Psycho-education Booklet for Genetic Aortic Disorders

As discussed in Chapter 2, studies have reliably shown that patients with chronic illnesses would like information about the nature, process, treatment, prognosis, family risk (Rutten et al., 2005; Coulter, Entwistle, & Gilbert, 1999; Adams, Boulton, & Wason, 2009). The study booklet included those topic categories listed above and the participants’ feedback confirmed that the booklet content was appropriate, comprehensive and desired.

Lifestyle topics such as exercise and diet, and the emergency plan were reported by participants to be the topics of greatest importance, all being highly rated by patients who saw their usefulness and relevance to daily life. It has been reported that individuals with a chronic illness face multiple physical, psychological, social, and lifestyle issues (Livneh & Antonak, 2005). Therefore, easily accessible information about how a GA disorder may affect the individual across multiple domains is important in increasing awareness and providing information about where to find support. In addition, as it is known that non-adherence to medical treatment increases morbidity and mortality (McHorney, 2009), patients require the knowledge about their diagnosis, treatment, and self-care issues to make informed choices and to gain a sense of control over a chronic condition where major lifestyle decisions and changes would be lifelong.

The lifestyle sections, particularly the topic of lifestyle changes and FAQs were also highlighted as useful topics in the booklet by the clinicians. From the clinicians’ perspectives, the lifestyle information depicted in the booklet would be useful in reinforcing the relevant verbal information given during the consultation, when there
were often time constraints. Other topics rated positively by clinicians included the section on coping with a diagnosis, and the section on myths. Clinicians commented that patients could refer back to the information in their own time and seek clarification or support as needed. The patient group also rated the section about frequently asked questions (FAQs), and dispelling common myths/misconceptions as useful tools that would enhance their ability and confidence in communicating with their medical team.

The study findings also endorsed reports that patients highly valued basic/general information at the time of diagnosis and then more specific information over time (Rutten et al., 2005; Piredda & Marfoli, 2004). This was reflected by suggestions from both patient and clinicians for further information on specific topics to be provided at later stages, such as post-surgery care, longer term self-care, and genetic issues. These topics had been considered for inclusion in the booklet during its development and in review phases, but it was determined that the current content would maintain a balance of simple but informative initial information, in contrast to providing detailed information about any one subject. This is in line with the aim of providing an overview of the most relevant topics associated with a recent diagnosis of a GA disorder. Readers who would like to seek more specific and comprehensive information about the topics of interest are able to refer to suggestions in the booklet on where to seek further information.

The patient feedback on the topic of emotions and coping with stress was divergent. Patients either strongly opposed or strongly identified with the need to have the topic in the booklet. From the comments given in the patient questionnaire, which were supported
and clarified by patient interview responses, it appeared that there were two main reasons for the opposing responses. Those supporting this topic identified that acknowledging and addressing emotional health and ways of coping was important. Those less interested in the topic said that they preferred to adopt a practical-oriented coping stance and felt that the topic was not of personal relevance. There was no clear link between liking or disliking the topic and time since diagnosis. The clinician group did not have any specific comments about the usefulness or acceptability of the coping topic.

It has been reported that the recall of information is poor at the time of initial consultation or serious diagnosis (Baumgartner, 2011; Butow, Brindle, McConnell et al., 1998). It has been found that the initial emotional experience associated with the diagnosis of a chronic illness can impede information processing and learning, it has been suggested that health information can be gathered by support persons until the individual is more ready to process the information (Baumgartner, 2011). Therefore information may be better given in increments to assist with increasing information needs over time. The study booklet addresses these concerns, as the booklet would be readily available for both the patient and support person as needed, on an ongoing basis with the option of being directed to where to access further information.

Research on the psychosocial impact and coping with chronic illness, including individuals with heart disease, cancer, epilepsy, rheumatoid arthritis, and diabetes, has shown two main coping styles: engagement and disengagement (Livneh & Antonak, 2005). Engagement includes strategies such as emotional expression, acceptance, and
problem-solving, whereas disengagement includes responses such as denial, avoidance, and inaction (Compas et al. 1999; Connor-Smith et al., 2000; Carver & Connor-Smith, 2010). It is known that an individual’s use of coping strategies can change depending on the situation (Livneh & Antonak, 2005). It is suggested that problem-focused coping may be more effective when the situation is changeable and controllable, and emotion-focused coping may be more suitable when the situation is unchangeable and uncontrollable (Livneh & Martz, 2007). In addition, it is known that the effectiveness of coping relies on a balance between the external context and the stress management strategies used by the individual. More specifically, adaptive coping depends on a range of strategies that is used flexibly and is a combination of emotion-focused and problem-focused approaches (Livneh & Martz, 2007). Given that individuals with a GA disorder may face various and multiple medical issues at different time points, it may be that rating the importance of the topic of coping with stress was influenced by the patient’s coping strategies that were either active or dominant at the time of their participation in the current study. By acknowledging the possibility of strong responses with a list of common experiences, followed by practical tips for coping with stress, the booklet is designed to allow readers to interpret the material in a way that would enhance their preferred coping orientation and strategies at different time points.

Another important reason in support of the inclusion of the coping section in the booklet is that psycho-social interventions for individuals with a chronic illness are becoming more available and effective (Turner & Kelly, 2000). Parker et al. (1995) found that stress management interventions were effective in reducing helplessness, dependency and pain.
Emotional expression strategies including verbal disclosure and writing/journaling have been found to be effective in reducing distress, improving mood and aid in adjustment in individuals with chronic illnesses such as asthma, rheumatoid arthritis, and breast cancer (de Ridder, Geenen, Kuijer, & van Middendorp, 2008; Stanton et al., 2002; Wetherell et al. 2005). The booklet acknowledges possible emotional difficulties and supports the reader to seek support if required.

5.2.2 Readability and Clarity

The readability, accessibility, and acceptability factors affect the quality of an information source (Coulter & Ellins, 2006), and the overall quality and usefulness of an information source are influenced by factors such as the design format, and the use of figures and diagrams. The format of the booklet, in terms of readability, terminology use, design the use of figures and diagrams, and size and length of the booklet, were positively appraised by both the patient and clinician groups.

Entwistle et al. (1998) emphasised the importance of the use of appropriate language, terminology, style, and tone, and clearly stating the purpose of the psycho-education material. The clinicians surveyed commented that the booklet was easy to read, with the language and level depth of information pitched at an appropriate level for most patients. Feedback from the patient group showed that the language style of the booklet was considered appropriate in its tone, that the language was clear and concise, that the booklet used medical terminology appropriately and avoided the use of technical jargon. The information was regarded as following a logical sequence, was well-structured and
focused. The patient group reported no difficulties with the literacy or reading level of the booklet, and that the language used to explain medical terms was at a layperson’s level. It has been shown that the effective use of plain language that avoids being simplistic or patronising facilitates effective communication and patients’ comprehension of health information (Wicklund & Ramos, 2009). Ensuring that a health information resource is uses appropriate language is essential in the context of health literacy.

Health literacy can be defined as the capacity to apply a range of functional skills, including general literacy skills, in the context of healthcare, in order for individuals to be involved in and make decisions about their treatment (Coulter & Ellins, 2006). As discussed in Chapter 2, it is known that inadequate health literacy is associated with poorer health status, lower treatment adherence, lower levels of disease management and health-promoting behaviours, and poorer ability to communicate with healthcare professionals and less involvement in shared decision-making (Ad Hoc Committee on Health Literacy, 1999; Sihota & Lennard, 2004). Whereas, the provision of quality health information such as printed materials promotes health literacy by aiding in patient knowledge gain and information recall, communication with health professionals, and involvement in health care (Coulter & Ellins, 2006). A systematic review conducted by McPherson and colleagues (2001) showed that written information enhanced recall and knowledge in patients with cancer. Similarly, Whelan and colleagues (2001) found from their systematic review that the use of decision aids in cancer increased patient’s knowledge and involvement in decision-making.
While formal readability tools were not used in the evaluation in the current study, the feedback questionnaires for both the patient and clinician groups contain specific questions about a range of readability factors. The current study booklet was positively appraised by both the patient and clinician groups for its use of appropriate language, terminology, style, and tone, that enhanced readability and promoted health literacy. In addition, feedback from the patient and clinician groups showed that the graphics supported and enhanced the text in the study booklet. In particular, patients commented that the use of a real scan enhanced their understanding of the related text. As recommended by Doak et al. (1996), graphics that carry content enhance readability for low literacy readers. Standardised readability and literacy tools may be appropriate for a subsequent RCT of the study booklet.

The booklet introduction and patient’s story were used to clearly state the purpose of the booklet and set the tone of the booklet as patient-centred. The patient group feedback showed the appreciation of these sections. The A5 (pocket) size of the booklet was also commended by patients and clinicians. The booklet was also considered the right length, allowing a general overview to be given without complicated or technical details. These factors combined enhanced the practicality and readability of the booklet in order to achieve the appropriate literacy and comprehension demands for the target audience.
5.2.3 Psychological wellbeing

From the results of the DASS21, it can be seen that the patient group did not have clinically significant psychological issues in relation to depression, anxiety, and stress. Self-report from the patients also indicated that individuals did not seem to experience any adverse impact from reading the booklet. This supported findings from other studies that reported no adverse anxiety or depression effects from health information exposure (Kennedy, Robinson, Hann, Thompson, & Wilkin, 2003; O'Brien, Whelan, Villasis-Keever, Gafni, Charles, Roberts, et al., 2009; Whelan, Levine, Willan, Gafni, Sanders, Mirsky et al., 2004). It has also been shown that patients prefer information whether it is good or bad news (Jenkins, Fallowfield, & Saul, 2001). However, it is not known whether individuals with pre-existing psychological concerns may experience a negative (short or long-term) impact from exposure to the booklet. A RCT with a larger sample is needed to ascertain a potential (positive or negative) psychological impact of this information resource across time.

5.2.4 Family impact

Given that GA disorders have a familial component, it was important to obtain feedback about the possible impact of the booklet on the patient’s family or other support persons. The patient interviews explored this issue, and showed that the booklet was perceived by patients as a valuable resource for family members and friends. Namely, the booklet could be used as a conversation starting point and provide an explanation about different aspects of the condition, treatment, and the issues discussed in appointments. In a systematic review of studies exploring the dynamics between patient, physician and
companions in communication and medical decision-making (Laidsaar-Powell, Butow, Bu, Charles, Gafni et al., 2013), it was found that companions often have a role in treatment decision-making outside of medical consultations. The study booklet would be beneficial as an information resource that both the patient and companion can refer to in their discussions away from the consultation.

It is also important to consider the companion support role within the medical consultation. In a systematic review by Laidsaar-Powell et al. (2013), it was reported that companions of patients with chronic illnesses most often played the primary role of emotional and informational support, including with decision-making, and communication with the medical team. In addition, through semi-structured interviews of oncology health professionals’ observations of family member roles in consultations, Laidsaar-Powell, Butow, Bu, Fisher, and Juraskova (2016) found that family members could also fulfil the roles of the devil’s advocate, apprentice healthcare provider, family liaison, and post-consultation support roles. Further, family involvement in cancer treatment decision-making has been reported to be variable throughout the course of treatment, starting before the initial medical consultation and continuing after the consultation (Laidsaar-Powell et al., 2016). It is therefore important to be aware that patients and support person’s needs and roles are varied and complex and may change over time, hence information provision needs to be adaptable. The current study booklet is flexible enough to be tailored or personalised in that relevant information can be highlighted by the clinician and extra information can be added as necessary.
Patients diagnosed with cancer reported that companion’s involvement were greatest at the beginning of treatment with the discussion of test results and decision-making (Beisecker, Brecheisen, Ashworth, & Hayes, 1997). When companions were involved in the medical consultation, patients reported that their understanding of the medical condition increased (Rosland, Piette, Choi & Heisler, 2011), and that companions helped with providing and eliciting information in consultations (Hubbard, Illingworth, Rowa-Dewar, Forbat, & Kearney, 2010). In addition, Clayman, Roter, Wissow and Bandeen-Roche (2005) found that when companions prompted the patient, asked the patient questions, and asked for the patient’s opinion in medical consultations, the companions were more likely to be actively involved in decision-making in comparison to companions who were not involved in that way. The current study’s booklet can serve as a reliable information source and may facilitate communication between the patient, support person(s) and clinician.

An important related issue is that it is known that carers of patients with cancer have many unmet needs themselves (Soothill, Morris, Harman, Francis, Thomas, & McIllmurray, 2001). The main unmet needs in caregivers of cancer patients are reported to include health care service and information needs, followed by emotional and psychological needs (Sklenarova et al., 2015). In a systematic review by Adams, Boulton, and Watson (2009), it was identified that the unmet needs of individuals affected by cancer, their partners, and family members, included diagnosis and prognosis related information, impact on the family, impact on the partner relationship, practical issues, coping, and treatment-related information. The booklet may be of benefit to family
members as a tool to aid in their understanding of GA disorders and the issues that the patient may face. In addition, it may be useful for family members who may consider seeking medical consultation or genetic testing for themselves. It was evident that the booklet had a broader reach and that accessibility of the booklet to family members would be valuable.

Summary

The development and evaluation of a psycho-education booklet for individuals diagnosed with a GA disorder described in this thesis was strongly endorsed by patients and clinicians as a valuable information source. The booklet was regarded positively in terms of its readability, content, format, and acceptability. The content of the booklet, particularly the sections on GA disorders, treatment options, and lifestyle changes was considered by both the patient and clinician groups overall to provide a comprehensive yet easy to read mix of factual/medical, practical, and person-centred information. The patient group indicated that the topics of greatest importance to them were: emergency planning, lifestyle issues, and further information sources. The QPL and FAQs were particularly valued by both the patient and clinician groups. Clinicians suggested that the booklet would form an important part of information giving at early diagnosis as the content gave a comprehensive overview of multiple topics related to a diagnosis to a GA disorder and may continue to be useful over time. The booklet was considered to meet its objectives in relation to its usefulness, clarity, and acceptability for individuals recently diagnosed with a GA disorder.
5.3 Limitations and strengths of the study

A limitation of this pilot study was the relatively small number of participants in the patient survey and interviews, which has implications for generalisability of the findings. The number of clinicians who specialise in the treatment of GA disorders is relatively limited, which was a barrier to recruitment for the clinician sample. Additionally, this study was conducted in one state (New South Wales) and therefore all patients recruited were attendees of the main clinic that specialises in the treatment of GA disorders and were consulting with the same specialist clinician. However, patients are referred to this clinic from a wide geographical area within the state.

Although the sample size in the current study was relatively small, it was appropriate for the purpose of conducting a pilot evaluation of the booklet. As this was a pilot study and GA disorders are rare conditions, a greater patient sample size was not required or feasible. The demographics and clinical/medical characteristics of the sample in the study represented a range of individuals affect by a GA disorder which reflects the variance seen in clinical practice.

As both the patient and clinician groups were asked to mail the completed questionnaires back to the researcher, this may have further hampered the response rate. All patients were English speaking and generally well-educated. These factors may have played a role in patients’ willingness to complete the study questionnaires and/or agree to an interview due to self-selection bias. The clinician sample consisted of cardiology specialists with many years of experience in this field. Responses from clinicians earlier in their career
and with a wider variety of medical specialties may differ from our results. Time constraints appeared to be one of the most likely factors in the relatively low response rate in the clinician sample.

The information from the questionnaires and responses from the patient interviews were reviewed as they became available. Therefore, the researchers actively implemented preliminary analysis of the trends and detected qualitative themes as they emerged over time. Despite the relatively small sample size in this study, theoretical saturation of the data was deemed to have been reached as no new trends/sub-themes developed and additional data would unlikely reveal new or further information.

Other pilot studies in the medical field, including oncology, cardiology, respiratory and primary care, have reported a variable range in sample size. Pilot studies that evaluated written health information resources or decision aids have reported similar sample sizes to the current study and yield important data that can guide the implementation of a RCT. For example, a pilot evaluation of a treatment decision aid for women with breast cancer utilised a patient sample size of \( n=17 \), and clinicians sample size of \( n=7 \) (Chiew, Shepherd, Vardy, Tattersall, Butow, & Leighl, 2008), and found that the majority rated the decision aid as acceptable, clear, and informative. An evaluation of an online decision aid for patients with osteoarthritis of the knee with a patient sample size of \( n=12 \) reported that participants rated the decision aid as easy to understand and user friendly (Washington, & Shacklady, 2014).
Lancaster and colleagues (2004) suggested that the testing of participant questionnaires is an important function of a pilot trial in order to ensure that the questionnaire is clear, comprehensible, and appropriate. This is particularly important for forms that are self-administered, as were the questionnaires utilised in the present study. Furthermore, Thabane et al. (2010) suggested that the objectives of pilot trials can include generating sample size calculations, and increase the likelihood of success in the RCT. It is hoped that the pilot study will facilitate the successful implementation of a RCT of the booklet by pre-testing the use of the study instruments and determining procedural strengths and barriers. It is evident that a greater response rate may be difficult to obtain in the naturally small participant pool in Australia, and that a RCT would need to expand the number of sites and/or reach out to an international audience.

A possible limitation of the study was that the booklet was not evaluated for its readability level using a structured readability tool nor was the patient sample assessed for health literacy using formal readability assessments. The readability of the booklet was however appraised through direct questions on a variety of aspects of the booklet’s design and use of language in the feedback questionnaire and there were opportunities for participants to make qualitative comments about the readability of the booklet. All the participants responded with “Agree/Strongly Agree” to readability statements such as: *The booklet was easy to understand; The booklet was easy to read,* and there were no other comments made by the participants on this topic. It may have been that the health literacy levels of the patient sample were functionally high as all spoke English as the primary language at home, and the sample were relatively well-educated. In a
subsequent RCT of the booklet, assessment of the health literacy of the patient sample could be incorporated and would be important as the sample would also be more diverse than the pilot sample.

Culturally and linguistically diverse (CALD) patients were under-represented in the current study since it was a study requirement that participants came from an English-speaking background (inclusion criteria was sufficient command of the English language to complete questionnaires / participate in an interview). It is acknowledged by the researchers that this booklet is unintended for and not suitable for a non-English speaking audience. Consequently, the experience of CALD patients with GA disorders remains an area for future investigation. It is possible that cultural and language barriers may prevent CALD patients from seeking help, or from understanding health information provided. A study examining the communication differences in oncology consultations with CALD cancer patients with or without interpreters in comparison to Anglo-Australians, found differences in interactions such as less time focused on cancer issues, and less summarising and informing, which the authors suggested may have an effect on decision-making (Butow et al., 2011).

The strength of this study was that the development of the booklet followed a structured process guided by standardised principles, adapted from O’Connor & Jacobsen (2003) and Coulter et al. (1999), which strengthened the reliability, rigour, and quality of the booklet. A needs assessment (Connors et al., 2015) was reviewed and updated, followed by an iterative process of drafting and review of the booklet. In addition, the
comprehensive pilot evaluation phase of the study consisted of a mixed methodology which obtained feedback from the patient population in the form of questionnaires and interviews, in addition to feedback from clinicians. Conducting feedback interviews gave opportunities to clarify discrepant or counter-intuitive findings reported in the questionnaires. The use of triangulation facilitated the validation of the data as it enabled the cross-verification from two different sources. This provided both qualitative and quantitative information which enabled the elucidation of complementary and divergent aspects of the data (Patton, 1999; Creswell, 2012). This triangulation of feedback from multiple stakeholders yielded valuable information about the content, format, utility, and acceptability of the booklet and ensured its ecological validity.

5.4 Future research

The results of the pilot evaluation described in this thesis strongly demonstrated the utility of the booklet as an important and appropriate psycho-education resource for individuals with a GA disorder. The next step in validating and furthering the utility of the booklet would be to conduct a RCT, followed by a clear booklet dissemination plan (if found effective). The effectiveness of the booklet in terms of knowledge gain and retention would be important outcome measures. It may be shown that the booklet promotes better understanding of the condition and treatment leading to increased coping and less distress (such as anxiety) than for patients who do not receive the booklet at diagnosis. Another aspect would be to assess medical adherence and self-management due to the knowledge gain in the immediate/short-term and longitudinally. Similarly to the current study, utilising quantitative and qualitative strategies could be useful in
examining these outcomes. Evaluations of other health resources has shown that patient education and information can improve coping, reduce anxiety, promote self-care and participation, and improve treatment adherence (Galloway et al., 1997; Mills and Sullivan, 1999; Treacy and Mayer, 2000; Chelf et al., 2001).

In addition, expanding the patient and clinician sample to include interstate or international participants would allow for a greater generalisability of findings. This could be achieved through a methodology utilising online and social media resources.

5.5 Clinical implications

In terms of clinical significance, the GA disorders psycho-education booklet has the potential to positively impact on patients’ understanding of medical issues, short/long term lifestyle changes and adherence to the chosen management option. In a qualitative study exploring the process of adjustment in patients with a GA disorder, Connors et al. (2015) found that patients wanted GA disorder-specific information resources that were provided by and endorsed by specialist clinicians. It was particularly important that the information resource would help the patient understand and navigate the treatment processes. It is also known that individuals with MFS can experience social isolation, psychological distress, reduced functioning and decreased overall quality of life (Fusar-Poli et al., 2008; Peters et al., 2005). The booklet can play an important part in supporting the individual and support persons to manage these psycho-social impacts. Additionally, the booklet can accommodate the patients’ preference for information, decision-making, and involvement in treatment planning, and clinicians have the option to verbally
highlight relevant topics in the booklet. Treatment non-adherence in chronic conditions is a known issue (DiMatteo, 2004), which impacts on morbidity and mortality (McHorney, 2009). Long-term self-management is required for patients with a GA disorder or other chronic condition, which can be influenced by improved patient-clinician communication and patient education (Brunton, 2011).

The results of the evaluation showed that the booklet was considered high in acceptability and usability by both the patient and clinician groups. Patients valued the booklet as an easily accessible information source and clinicians endorsed its use. The booklet could be routinely provided to patients with a GA disorder at the time of diagnosis or early treatment. Patients would then have the option to refer to the booklet as and when needed, during or between consultations over time. The booklet could also facilitate or enhance the communication between patient and clinician, and/or support persons. Furthermore, patients could be encouraged to use the booklet as a way to provide information to partners, family and friends.

The booklet in printed form is a low-cost and portable resource. Its accessibility could be expanded for little or no cost by being published online. As participants requested additional information about surgical and lifestyle topics, there is a need for the development and evaluation of resources relating to those issues. Those additional resources could also be made available through links within the online version of the booklet. Translation of the booklet into other languages would also broaden its reach.
5.6 Conclusion

This thesis describes the development and pilot evaluation of an evidence-based psycho-education booklet for patients recently diagnosed with a GA disorder. The main objectives of the pilot evaluation of the booklet were to assess the booklet’s usefulness, clarity, and acceptability by the patient and clinician population.

The development of the psycho-educational booklet addresses the unmet information needs of individuals with a GA disorder. The results of the evaluation showed that the study booklet was considered a highly appropriate, practical, informative, and useful psycho-education resource by both patients and clinicians who participated in this study. To our knowledge, this will be the first GA disorders psycho-education booklet developed and pilot evaluated specifically for an Australian audience.
References


The Development and Evaluation of a Psycho-education Booklet for Genetic Aortic Disorders


Harris, J. (1992). You can't ask if you don't know what to ask: a survey of the information needs and resources of hospital outpatients. *The New Zealand medical journal, 105*(934), 199.


The Development and Evaluation of a Psycho-education Booklet for Genetic Aortic Disorders


The Development and Evaluation of a Psycho-education Booklet for Genetic Aortic Disorders


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The Development and Evaluation of a Psycho-education Booklet for Genetic Aortic Disorders


The Development and Evaluation of a Psycho-education Booklet for Genetic Aortic Disorders


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The Development and Evaluation of a Psycho-education Booklet for Genetic Aortic Disorders


Appendices

Appendix A  Lead Human Research Ethics Committee Approval
Appendix B  Royal Prince Alfred Hospital Site Specific Approval
Appendix C  Patient Invitation Letter
Appendix D  Patient Information Form
Appendix E  Patient Consent Form
Appendix F  Patient Feedback Questionnaire
Appendix G  Patient Interview Schedule
Appendix H  Clinician Information Letter
Appendix I  Clinician Feedback Questionnaire
Appendix J  Booklet: Genetic Aortic Disorders
Appendix A  Lead Human Research Ethics Committee Approval

7 May 2013

Dr I Juraskova
School of Psychology
CeMPED
Brennan MacCallum Building, A18
UNIVERSITY OF SYDNEY NSW 2006

Dear Dr Juraskova,


The Executive of the Ethics Review Committee, at its meeting of 2 May 2013, considered your correspondence of 18 April 2013. In accordance with the decision made by the Ethics Review Committee, at its meeting of 13 March 2013, ethical approval is granted.

The proposal meets the requirements of the National Statement on Ethical Conduct in Human Research.

This approval includes the following:

- NEAF (AU/1/1361110)
- Participant Invitation Letter (Patients) (Master Version 2.0, 18 April 2013)
- Information for Participants (Patients) (Master Version 2.0, 18 April 2013)
- Participant Consent Form (Master Version 1.0, 12 February 2013)
- Participant Questionnaire (Patients) (Master Version 2.0, 18 April 2013)
The Development and Evaluation of a Psycho-education Booklet for Genetic Aortic Disorders

- Participant Invitation Letter (Clinician) (Master Version 1, 14 February 2013)
- Information for Participants (Clinicians) (Master Version 2.0, 18 April 2013)
- Clinician Online Questionnaire (dated 19 April 2013)

You are asked to note the following:

- This letter constitutes ethical approval only. You must NOT commence this research project at ANY site until you have submitted a Site Specific Assessment Form to the Research Governance Officer and received separate authorisation from the Chief Executive or delegate of that site.

On the basis of this ethics approval, authorisation may be sought to conduct this study within any NSW public health organisation and/or within any private organisation which has entered into an appropriate memorandum of understanding with the Sydney Local Health District, Sydney Local Health Network or the Sydney South West Area Health Service.

The Committee noted that authorisation will be sought to conduct the study at the following sites:

- Royal Prince Alfred Hospital

This approval is valid for four years, and the Committee requires that you furnish it with annual reports on the study's progress beginning in June 2014. If recruitment is ongoing at the conclusion of the four year approval period, a full re-submission will be required. Ethics approval will continue during the re-approval process.

This human research ethics committee (HREC) has been accredited by the NSW Department of Health as a lead HREC under the model for single ethical and scientific review and is constituted and operates in accordance with the National Health and Medical Research Council's National Statement on Ethical Conduct in Human Research and the CPMP/ICH Note for Guidance on Good Clinical Practice.

- You must immediately report anything which might warrant review of ethical approval of the project in the specified format, including unforeseen events that might affect continued ethical acceptability of the project.

- You must notify the HREC of proposed changes to the research protocol or conduct of the research in the specified format.

- You must notify the HREC and other participating sites, giving reasons, if the project is discontinued at a site before the expected date of completion.

- If you or any of your co-investigators are University of Sydney employees or have a conjoint appointment, you are responsible for informing the University's Risk Management Office of this approval, so that you can be appropriately indemnified.
Where appropriate, the Committee recommends that you consult with your Medical Defence Union to ensure that you are adequately covered for the purposes of conducting this study.

Should you have any queries about the Committee's consideration of your project, please contact me. The Committee's Terms of Reference, Standard Operating Procedures, membership and standard forms are available from the Sydney Local Health District website.

A copy of this letter must be forwarded to all site investigators for submission to the relevant Research Governance Officer.

The Ethics Review Committee wishes you every success in your research.

Yours sincerely,

Lesley Townsend
Executive Officer
Ethics Review Committee (RPAH Zone)

HERC\EXCOR\13-05
Appendix B  Royal Prince Alfred Hospital Site Specific Approval

1 August 2013

Professor J Richmond
C/- Dr I Juraskova
School of Psychology (CeMPED)
Room 160, Transient Building (F12)
UNIVERSITY OF SYDNEY NSW 2006

Dear Professor Richmond

Re: Protocol No X13-0204 - “A randomised controlled trial of an information booklet on Genetic Aortic Disorders”

HREC/13/RPAH/243  SSA/13/RPAH/341

Thank you for submitting a Site Specific Assessment Form for this study. I am pleased to inform you that authorisation has been granted for it to be undertaken at the Royal Prince Alfred Hospital Marfan and Aortic Diseases Clinic.

The approved information and consent documents for use at this site are:

- Participant Invitation Letter (Version 2.0, 25 June 2013)
- Information for Participants – phase 2 (Version 2, 25 June 2013)
- Participant Consent Form (Version 2, 25 June 2013)
- Participant Letter for Follow-up Questionnaire (Version 2, 25 June 2013)
- Participant Reminder Letter (Version 2, 25 June 2013)

The following conditions apply to this research study. These are additional to those conditions imposed by the human research ethics committee (HREC) that granted ethical approval:

1. Proposed amendments to the research protocol or conduct of the research, which may affect the ethical acceptability of the study and which are submitted to the lead HREC for review, must be copied to me.
2. Proposed amendments to the research protocol or conduct of the research, which may affect the ongoing site acceptability of the study, must be submitted to me.

I wish you every success in your research.

Yours sincerely,

Lesby Townsend
Research Governance Officer
SSWAHS (RPAH Zone)
RGO - LesbyCORRESP12-0207
Appendix C  Patient Invitation Letter

We would like to let you know about a research study evaluating the usefulness of an information booklet on genetic aortic disorders. The aim is to improve the experiences and resources for people recently diagnosed with a genetic aortic disorder, and specifically we would like to get feedback on a newly developed booklet. To our knowledge, there have been no studies looking at the use of an information booklet about genetic aortic disorders in Australia. Any feedback you may give us will help us provide better support and resources to other people in a similar situation. This is a collaborative project between the Marfan and Aortic Disease Clinic at the Royal Prince Alfred Hospital (RPAH) and the University of Sydney.

Participation in this study is voluntary, and will have no impact on the level of care you receive. Your treating team will not be aware of whether or not you decide to participate in this study, and you can withdraw from the study at any time, without giving a reason. Although the research may not directly benefit you, it will help to improve the information booklet which will be made available to people diagnosed with a genetic aortic disorder in the future.

Enclosed in this package is a Participant Information Sheet, Consent Form, the Information Booklet, Study questionnaire, and pre-paid self-addressed envelope.

Participation in the study involves reading the booklet and completing the enclosed questionnaire. The questionnaire will take approximately 30-40 minutes of your time, and provide us with information about how to improve the content and presentation of the information booklet.

If you agree to take part in this study, please return:

1. the signed consent form
2. the completed questionnaire

in the reply paid envelope provided. If you do not wish to participate, please dispose of this study package in whichever way you choose.

If you have any questions, please contact Dr Ilona Juraskova or Ms Joycelyn Ling at any time on (02) 9036 5275, or Professor Richmond Jeremy on (02) 9515 8766.

Thank you for considering this invitation.
Yours sincerely

[Signature]

Richmond Jeremy
Appendix D  Patient Information Form

Evaluation of an Information Booklet on Genetic Aortic Disorders

INFORMATION FOR PARTICIPANTS

Introduction
You are invited to take part in a research study into the evaluation of an information booklet on genetic aortic disorders. The aim is to assess the booklet’s readability, clarity, and usefulness for people who have recently been diagnosed with a genetic aortic disorder.

The study is being conducted by Joycelyn Ling and will form the basis for the degree of Doctor of Clinical Psychology/Master of Science at the University of Sydney under the supervision of Dr. Ilona Juraskova (The University of Sydney) and Professor Richmond Jeremy (The University of Sydney/The Sydney Heart Clinic, RPAH).

Study Procedures
If you agree to participate in this study, you will be asked to sign the Participant Consent Form, read the enclosed booklet, complete the included Questionnaire, and return the signed Consent Form and completed Questionnaire to the researcher.

The information being sought in the questionnaire includes: your view on the presentation and content of the information in the booklet; your knowledge relating to genetic aortic disorders; psychological wellbeing.; and your typical decision-making style.

We would also welcome verbal feedback on the booklet and the knowledge scale. If you agree to participate in a telephone interview, as indicated by completing the Contact Form, the researcher will contact you to arrange a time to conduct a phone interview. The consent form, reading the booklet, and completing the questionnaire will take approximately 45-60 minutes of your time. The telephone interview will take approximately 20-30 minutes.

Risks
It is possible that you may experience some distress in reading the information booklet and completing the questionnaire. Should you become distressed you have the option to withdraw from the study. Dr. Ilona Juraskova (research supervisor) can also be contacted on (02) 9036 5275. If needed, you may also be referred to an appropriate support service.

Benefits
Although the research may not directly benefit you, it will help to improve the information booklet and in developing other support resources for people with genetic aortic disorders.

Costs
Participation in this study will not cost you anything, nor will you be paid.
Voluntary Participation
Participation in this study is entirely voluntary. You do not have to take part in it. If you do take part, you can withdraw at any time without having to give a reason. Whatever your decision, please be assured that it will not affect your medical treatment or your relationship with the staff who are caring for you.

Confidentiality
All the information collected from you for the study will be treated confidentially, and only the researchers named above will have access to it. The study results may be presented at a conference or in a scientific publication, but individual participants will not be identifiable in such a presentation.

Further Information
When you have read this information, Joycelyn Ling or Ilona Juraskova will discuss it with you further and answer any questions you may have. If you would like to know more at any stage, please feel free to make contact on (02) 9036 5275.

Ethics Approval and Complaints
This study has been approved by the Ethics Review Committee (RPAH Zone) of the Sydney Local Health District. Any person with concerns or complaints about the conduct of this study should contact the Executive Officer on 02 9515 6766 and quote protocol number X13-0061.

This information sheet is for you to keep.
Appendix E  Patient Consent Form

Evaluation of an Information Booklet on Genetic Aortic Disorders

PARTICIPANT CONSENT FORM

I, ..........................................................................................................

[name]

Of ..........................................................................................................

[address]

have read and understood the Information for Participants on the above named research study.

I have been made aware of the procedures involved in the study, including any known or expected inconvenience, risk, discomfort or potential side effect and of their implications as far as they are currently known by the researchers.

I freely choose to participate in this study and understand that I can withdraw at any time.

I also understand that the research study is strictly confidential.

I hereby agree to participate in this research study.

NAME:

..........................................................................................................

CONTACT No.: (Home) ..........................................................................

(Mobile) ..........................................................................................

SIGNATURE:

..........................................................................................................

DATE:

..........................................................................................................

NAME OF WITNESS:

..........................................................................................................

SIGNATURE OF WITNESS:

...........................................................................................................
Appendix F  Patient Feedback Questionnaire

SECTION 1: DEMOGRAPHICS AND MEDICAL HISTORY
This section of the questionnaire asks some general questions about you. Please complete the field or tick (✓) the statement that best describes you.

Today’s date: _______________

What is your date of birth? _______________

Your gender

☐ Male
☐ Female

What is your present marital status?
Never married
Married / De facto
Widowed
Divorced / Separated

Do you have any children? If so, how many?

What is the highest education qualification you obtained?
Year 10 or below
Year 12 / HSC or leaving certificate
TAFE certificate / diploma
University degree
Higher degree (postgraduate)

What is your occupation (or previous occupation if retired)?

In which country were you born?
☐ Australia
☐ Other _____________________________

Do you speak a language other than English at home?
No, only English
Yes, I speak _____________________________

(Please specify)
The Development and Evaluation of a Psycho-education Booklet for Genetic Aortic Disorders

This section of the questionnaire asks some general questions about your diagnosis. Please complete the field or tick (✔) the statement that best describes you.

When were you diagnosed with a Genetic Aortic Disorder? ________________
(dd/mm/yyyy)

Have you had any surgical operations as a result of your condition?
No
Yes, Stent Graft
Yes, Valve Replacement
Other (Please specify) __________________________________________________________________

If yes, was your operation preventative or emergency?
Preventative
Emergency

Which information source about your condition have you accessed (if any)?
☐ Health or medical related internet website. Please specify
☐ Internet website about genetic aortic disorders. Which one(s):
☐ Book. Which one(s):
☐ Information sheets. Which one(s):
☐ From someone else. Who:
☐ Other. Please give details:

Some people prefer to have very few details about their illness while others prefer to have as many details as possible.
Please place a cross (X) on the line below to indicate your preference for information, on a scale of 0 (prefer as few details as possible) to 10 (prefer as many details as possible).

Prefer as few details as possible | 0 | Prefer as many details as possible
SECTION 3: FEEDBACK ABOUT THE BOOKLET

This section of the questionnaire asks some questions about the information in the booklet and the way that it is presented. Indicate how much you agree or disagree with each statement by circling the number that fits your response.

<table>
<thead>
<tr>
<th>Statement</th>
<th>Strongly Disagree</th>
<th>Disagree</th>
<th>Not Sure</th>
<th>Agree</th>
<th>Strongly Agree</th>
</tr>
</thead>
<tbody>
<tr>
<td>The booklet was easy to read</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>4</td>
<td>5</td>
</tr>
<tr>
<td>The booklet was easy to understand</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>4</td>
<td>5</td>
</tr>
<tr>
<td>I liked the look of the booklet</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>4</td>
<td>5</td>
</tr>
<tr>
<td>The booklet was the right length</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>4</td>
<td>5</td>
</tr>
<tr>
<td>The booklet was helpful to me overall</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>4</td>
<td>5</td>
</tr>
<tr>
<td>I felt anxious while reading the booklet</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>4</td>
<td>5</td>
</tr>
<tr>
<td>The booklet answered questions about how my condition may affect me</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>4</td>
<td>5</td>
</tr>
<tr>
<td>After reading the booklet, I know more about what to expect with treatment</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>4</td>
<td>5</td>
</tr>
<tr>
<td>The booklet provided information on how to best care for myself</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>4</td>
<td>5</td>
</tr>
<tr>
<td>The section on Coping with the emotional ups and downs (p.17) was useful</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>4</td>
<td>5</td>
</tr>
<tr>
<td>I would bring and refer to this booklet in future consultations with my physician</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>4</td>
<td>5</td>
</tr>
<tr>
<td>I would recommend this booklet to other people with a genetic aortic disorder</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>4</td>
<td>5</td>
</tr>
</tbody>
</table>

The information of most importance to me were: (Please indicate why)

The information of least or no importance to me were: (Please indicate why)

Do you have any other comments about the booklet?
SECTION 4: PSYCHOLOGICAL WELL-BEING
This section of the questionnaire asks some questions about whether you are experiencing stress, anxiety, or depression. Please read each statement and circle a number 0, 1, 2, or 3 which indicates how much the statement applied to you over the past week.

<table>
<thead>
<tr>
<th>DASS21</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>The rating scale is as follows:</td>
<td></td>
</tr>
<tr>
<td>0  Did not apply to me at all</td>
<td></td>
</tr>
<tr>
<td>1  Applied to me to some degree, or some of the time</td>
<td></td>
</tr>
<tr>
<td>2  Applied to me to a considerable degree, or a good part of time</td>
<td></td>
</tr>
<tr>
<td>3  Applied to me very much, or most of the time</td>
<td></td>
</tr>
<tr>
<td>1</td>
<td>I found it hard to wind down</td>
</tr>
<tr>
<td>2</td>
<td>I was aware of dryness of my mouth</td>
</tr>
<tr>
<td>3</td>
<td>I couldn't seem to experience any positive feeling at all</td>
</tr>
<tr>
<td>4</td>
<td>I experienced breathing difficulty (eg, excessively rapid breathing, breathlessness in the absence of physical exertion)</td>
</tr>
<tr>
<td>5</td>
<td>I found it difficult to work up the initiative to do things</td>
</tr>
<tr>
<td>6</td>
<td>I tended to over-react to situations</td>
</tr>
<tr>
<td>7</td>
<td>I experienced trembling (eg, in the hands)</td>
</tr>
<tr>
<td>8</td>
<td>I felt that I was using a lot of nervous energy</td>
</tr>
<tr>
<td>9</td>
<td>I was worried about situations in which I might panic and make a fool of myself</td>
</tr>
<tr>
<td>10</td>
<td>I felt that I had nothing to look forward to</td>
</tr>
<tr>
<td>11</td>
<td>I found myself getting agitated</td>
</tr>
<tr>
<td>12</td>
<td>I found it difficult to relax</td>
</tr>
<tr>
<td>13</td>
<td>I felt down-hearted and blue</td>
</tr>
<tr>
<td>14</td>
<td>I was intolerant of anything that kept me from getting on with what I was doing</td>
</tr>
<tr>
<td>15</td>
<td>I felt I was close to panic</td>
</tr>
<tr>
<td>16</td>
<td>I was unable to become enthusiastic about anything</td>
</tr>
<tr>
<td>17</td>
<td>I felt I wasn't worth much as a person</td>
</tr>
<tr>
<td>18</td>
<td>I felt that I was rather touchy</td>
</tr>
<tr>
<td>19</td>
<td>I was aware of the action of my heart in the absence of physical exertion (eg, sense of heart rate increase, heart missing a beat)</td>
</tr>
<tr>
<td>20</td>
<td>I felt scared without any good reason</td>
</tr>
<tr>
<td>21</td>
<td>I felt that life was meaningless</td>
</tr>
</tbody>
</table>
Appendix G  Patient Interview Schedule

**Question 1: Initial Response**
What were your first impressions of this booklet?
What did you like about it?  What was the best part?
What did you dislike about it?
Overall do you think a booklet like this is useful for a person who has recently received a diagnosis of a Genetic Aortic disorder? Why…?

**Question 2: Layout**
What did you think about the ‘look’ or ‘design’ of the booklet, e.g., the colours; the size of the writing; anything about the way it is presented?
What do you think about the pictures?
What did you think about the length of the booklet?  Was it:
  - Too long  [ ]
  - The right length  [ ]
  - Too short  [ ]
Was there:
  - Too much information  [ ]
  - The right amount of information  [ ]
  - Not enough information  [ ]

**Question 3: Timing**
Based on your experience, when do you feel would be the best time to receive the booklet?
Would you prefer if a member of your treatment team went through some of the pages with you before you took the booklet home?  If so, which ones?

**Question 4: Now we will focus on specific sections of the booklet**
Was the introduction of the booklet clear in describing the purpose of the booklet?
Was Josh’s story of interest to you? Why/why not?
Were the information sections helpful? e.g. description of the types of GA disorders, about aneurysms, treatment, in an emergency, surgery.
Were the sections about lifestyle, FAQs, Myths & Misconceptions, Coping, and More information helpful?
What did you think about the sections at the end where the treatment plan, appointments, test results, questions, and notes can be written?

**Question 5**
Are there any topics or questions that you feel were not covered in the booklet that should be included?
Yes  [ ]  No  [ ]
If yes, please tell us what you think should be added.

Are there any topics or information that you think should not have been included in the booklet?
Yes  [ ]  No  [ ]
If yes, please tell us what you think should be removed
Question 6
This booklet may be helpful for other people around you. Did anyone else read through the booklet? (If yes, please say which apply)

Yes  [ ]  No  [ ]

- Spouse/Partner  [1]
- Child  [2]
- Other family  [3]
- Friend  [4]
- Other  [5]

If anyone else read the booklet, did they find it helpful?

Very helpful  [1]
A bit helpful  [2]
Not helpful  [3]
Not sure  [4]

Did the booklet make it easier to talk about what you are experiencing/have experienced?

Question 7
Do you have any other comments that you would like to make?
Appendix H Clinician Information Letter

Evaluation of an Information Booklet on Genetic Aortic Disorders

INFORMATION FOR PARTICIPANTS

Introduction
You are invited to take part in a research study into the evaluation of an information booklet on genetic aortic disorders. The aim is to assess the booklet’s readability, clarity, and usefulness for individuals with a recent diagnosis of a genetic aortic disorder, and to assess clinicians’ perspective about using the booklet as an adjunct means of information delivery, in terms of its usefulness, benefit, and acceptability. In addition, we would like your feedback about a Knowledge Scale we are developing to be used to assess patients’ level of knowledge about genetic aortic disorders.

The study is being conducted by Joycelyn Ling and will form the basis for the degree of Doctor of Clinical Psychology/Master of Science at the University of Sydney under the supervision of Dr. Ilona Juraskova (The University of Sydney) and Professor Richmond Jeremy (The University of Sydney/The Sydney Heart Clinic, RPAH).

What does participation in this study require
If you agree to participate in this study, you will be asked to read the Booklet and complete an anonymous online or paper questionnaire. This will take approximately 30-45 mins. The questionnaire will help us to obtain your view on the presentation and content of the information in the booklet, and feedback on the usefulness of the booklet.

Risks
There are no known risks to you from being involved in this study. If you have any concerns while taking part in the study, please contact the researchers on (02) 9036 5275.

Benefits
Although the research may not directly benefit you, it will contribute to furthering the development of the information booklet. The findings may assist in guiding future research on the information needs and preferences of people recently diagnosed with a genetic aortic disorder, and for the development of other information and educational tools.

Costs
Participation in this study will not cost you anything, nor will you be paid.

Voluntary Participation and Consent to Participate
This questionnaire is anonymous. Your participation in the study is entirely voluntary and you are under no obligation to consent to participate. If you do consent to participate, you can withdraw your consent at any point while you complete the survey by disconnecting from the website or discarding the questionnaire. However, once the survey has been submitted by clicking the submit button, your data will not be able to be withdrawn as your responses cannot be identified for removal.
Clicking on "Yes" to “Take the Survey?” (online survey) indicates that, having read the information provided, you have understood the purpose and risks of the study. Your consent to participate in the survey for this project will be implied by your submission of the questionnaire by clicking the “Submit” button at the end of the questionnaire or by returning the questionnaire to the researchers.

**Confidentiality**
All the information collected from you for the study will be treated confidentially, and only the researchers named above will have access to it. The study results may be presented at a conference or in a scientific publication, but individual participants will not be identifiable in such a presentation.

**Further Information**
If you would like to know more at any stage, please feel free to contact Joycey Ling or Ilona Juraskova on (02) 9036 5275.

**Ethics Approval and Complaints**
This study has been approved by the Ethics Review Committee (RPAH Zone) of the Sydney Local Health District. Any person with concerns or complaints about the conduct of this study should contact the Executive Officer on 02 9515 6766 and quote protocol number X13-0061.

You may print a copy of the Participant Information Sheet for your records, should you so require.
Appendix I  Clinician Feedback Questionnaire

Question 1. This section of the questionnaire asks some general questions about you. Please complete the field or check the box of the statement that best describes you.

1a. Your gender
☒ Male
☒ Female

1b. What is your clinical field (and specialty, if relevant)?
☒ Cardiologist
☒ GP
☒ Surgeon
☒ Genetic counsellor
☒ Other, please specify ____________________

1c. How many years of experience do you have with people with GA disorders?

1d. Please describe your service type
☐ Public
☐ Private
☐ Community
☐ Other, please specify ____________________

1e. Most common Genetic Aortic Disorders diagnoses/conditions in your patient group (if relevant):

1f. Age range of patient group with Genetic Aortic Disorders

1g. Do you currently use patient information booklets or other educational material?
☒ Yes
☒ No

1h. If so, what type of material?

1i. If so, how often and when?

1j. If not, what are the reasons?
The Development and Evaluation of a Psycho-education Booklet for Genetic Aortic Disorders

Question 2. Your Initial Response to the Booklet
2a. What were your first impressions of this booklet?
2b. Overall, why do you think a booklet like this would be useful for a person who has recently (within 6 months) received a diagnosis of a GA disorder?
2c. Overall, why do you think a booklet like this may be useful as an information tool in consultations?

Question 3. Layout of the Booklet
3a. Please comment on the ‘look’ or ‘design’ of the booklet, e.g., the colours; the size of the writing; anything about the way it is presented?
3b. Please comment about the graphics (e.g. pictures, quote boxes, information highlight boxes etc.)

Question 4. Use of the Booklet
4a. Which patients do you think the booklet would be most suitable for?
4b. Which patients do you think the booklet would be unsuitable for?
4c. When do you think would be the best time to give patients the booklet?
4d. Should a member of the patient’s treatment team go through some of the pages with the patient before it was taken home?
   ☑ Yes
   ☑ No
4e. If so, who do you recommend this person should be?

Question 5. Content of the Booklet
5a. Are there any topics or questions that you feel were not covered in the booklet that should be included? If yes, please tell us what you think should be added:
5b. Are there any topics or information that you think should not have been included in the booklet? If yes, please tell us what you think should be removed:
5c. Do you have any other feedback or comments that you would like to make?
### Question 6. Feedback about the Booklet

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Please indicate how much you agree or disagree with each statement by clicking in the column that fits your response.
Appendix J  Booklet: Genetic Aortic Disorders
GENETIC AORTIC DISORDERS

Information about Genetic Aortic Disorders and Aortic Aneurysm
The purpose of this booklet is to give you and your family information about Genetic Aortic Disorders and their treatment, and to answer commonly asked questions.

This booklet can be brought to your appointments so that important information and reminders can be written down, and you can note down any questions that you may have in between appointments.

This booklet is not intended to replace direct medical information and advice from your treating physician. If you have any questions about your condition or the information in this booklet, please consult your physician for clarification.

"The more you know, the more power you have, and the better your control over your own health and well-being"

Professor Richmond Jeremy
Director, Marfan and Aortic Disease Clinic
Royal Prince Alfred Hospital
Sydney, Australia
How This Booklet Can Help You

You have most likely been given this booklet because you or a close family member has been diagnosed with a condition known as a Genetic Aortic Disorder.

Many people have questions about this condition, such as what it means to have it, what treatment may be recommended, and what impact it may have on their life and lifestyle. The information in this booklet has been designed to help answer these questions, and to point to further sources of information.

Josh’s Story

“I am 25 and was about to travel in Asia with my girlfriend. My family doctor said he heard a heart murmur and sent me for tests. Next thing I know, I’m seeing a heart specialist who says I have a serious problem with my aorta and I will need surgery soon. Then he said I can’t do weight training anymore. Where did this come from? Why me? I haven’t done anything to anybody…”

“My world was turned upside down…”

“My world was turned upside down for a while, but I was determined to find out as much information as I could about what I needed to do to recover after the surgery. I also talked to my doctor and my specialist about what I needed to do to stay healthy in the long term. Thinking about going on the trip with my girlfriend was the thing that kept me going on the down days. Guess what? We’ve just returned from an awesome 6 month trip!!!”
About Genetic Aortic Disorders

What is the Aorta?

The aorta is the main artery distributing blood from the heart to the body. It runs through the chest and abdomen and has numerous large branches. It is an elastic artery with walls made of many complex proteins.

What are Genetic Aortic Disorders?

Genetic Aortic (GA) disorders are inherited conditions which affect the protein structure of the aorta and sometimes also the valves inside the heart. These conditions are due to changes in the person’s genetic code, which are present in an individual from conception.

These conditions include Marfan Syndrome (MFS), Loeys-Dietz syndrome (LDS), Thoracic Aortic Aneurysm and Dissection (TAAD), and Bicuspid Aortic Valve Aneurysm Syndrome (BAVAS). An important consequence of these conditions is a tendency for the aorta to enlarge with time, leading to eventual tear (dissection) or rupture, which is life-threatening.

Who can be affected by a genetic aortic disorder?

Every human has variations or mutations in their genetic code (DNA). We are all individuals and the different DNA variations affect us all in their own way. Sometimes we inherit the DNA variations from our parents (autosomal inheritance) and sometimes the DNA variations result from small changes in copying the DNA code from our parents (de novo mutations).

Marfan Syndrome (MFS)

MFS affects the connective tissue of the body and impacts upon the skeleton, heart, eyes, lungs, blood vessels and nervous system. The condition occurs in approximately 1 out of 5,000 people and affects both males and females of any race or ethnic group. About 75 out of 100 affected individuals inherit the condition from one of their parents.

Loey-Dietz Syndrome (LDS)

LDS is rarer than MFS but has many similar physical symptoms to MFS, with other characteristics such as twisting or spiralled arteries in neck vessels, widely spaced eyes, split or broad uvula. Approximately 25 out of 100 cases of LDS are inherited.

Thoracic Aortic Aneurysm and Dissection (TAAD)

TAAD is usually asymptomatic and often has no external physical features. It may only be discovered by chance, or when aortic dissection or rupture occurs.

Bicuspid Aortic Valve Aneurysm Syndrome (BAVAS)

A bicuspid aortic valve has two instead of three leaflets, and lies between the heart and the aorta. It can be associated with weakening of the aortic wall, leading to an aneurysm forming. Approximately 1 in 100 people have a bicuspid aortic valve, of whom half will have some aortic dilation (enlargement). Approximately 30% of patients with BAVAS will have other affected family members.

We all have DNA variations (mutations)
DNA variations affect each of us in different ways

Normal aortic valve
Bicuspid aortic valve
What is an aortic aneurysm?

An aneurysm is a bulge (dilation) in the wall of an artery, usually in a weak area of the aorta. The pressure of blood inside the artery forces the weak area to balloon outward.

Symptoms of an aortic aneurysm may include chest or back pain. If untreated, an aneurysm may rupture, resulting in internal bleeding. These complications can be avoided through detection, surveillance, and surgical repair of the aneurysm.

Modern aortic surgery has excellent results and there is a wide variety of techniques, including Dacron implants, stenting, and valve repairs and replacements which can be tailored to suit individual needs.

What is an aortic dissection?

Aortic dissection occurs when the layers of the aorta tear and separate from each other, allowing blood to leak into the layers. The presence of an aortic aneurysm increases the risk of having an aortic dissection, but an aortic dissection can also occur in people with a normal sized aorta.

Aortic dissection is characterised by severe pain in the chest or back, and is a medical emergency.

Types of aortic aneurysms

Types of aortic dissection
How is an aortic aneurysm diagnosed?

Your physician may order the following tests to help with diagnosis:

**Chest x-ray** produces images of internal tissues, bones, and organs onto film.

**Computed tomography (CT) scan** uses X-rays to take images to produce cross-sectional and 3D pictures of the aorta. The images are used to examine the size and extent of an aneurysm or dissection. An injection of an iodine-based dye is typically given in this procedure.

**Magnetic resonance imaging (MRI)** produces cross-sectional and 3D images of the aorta, as well as blood flow in the aorta, by using a strong magnetic field and radio waves. An injection of a contrast dye is also used in this procedure. People with impaired kidney function or metal implants cannot undergo MRI.

**Angiography** is an x-ray of the blood vessels. An injection of a contrast dye is used in this procedure.

**Echocardiography**, or an ultrasound of the heart, is used to examine the heart, aortic valve, and aorta above the heart. The diameter of the aorta can be measured and compared, to monitor changes over time.

![Echocardiogram showing measurement of aortic aneurysm size](image)

Treatment: What to expect

Medical intervention for GA disorders involve life-long monitoring, and treatment of associated issues. The treatment of an aortic aneurysm depends on the presence of symptoms, geometry, and size, and will vary with individual circumstances. Treatment may include medication and surgery.

**Active Monitoring and Medication**

Close monitoring of the aorta is done with echocardiography every 6-12 months to detect changes. Regular monitoring of aortic size is critical to successful treatment.

Blood pressure medication may be required to lower the overall blood pressure and the pressure on the weakened area of the aneurysm. A cholesterol lowering medication may also be prescribed.

Your physician will discuss with you the purpose of the medication, dosage, side-effects, and long-term plan. If you have any urgent questions about your medication, contact your GP, or the clinic directly. Otherwise, space is provided on page 19 to write down questions for discussion with your physician at your next appointment.

The purpose of medication is to lower blood pressure and reduce stress on the abnormal aortic wall.
Surgical Repair

If the aortic aneurysm is at risk of rupture or is causing symptoms, surgery will be needed to prevent the rupture and to repair the weakened section of the aorta. Surgery may involve open repair, valve replacement, and stenting (where a synthetic fabric tube supported by a metal structure is inserted into the affected part of the aorta).

Decisions about the timing of surgery are made on an individual basis and there is time to prepare for a stay in hospital. Most people are only in hospital for about a week, and are back to their usual activities by approximately 6 weeks after surgery.

After surgery, normal activities can usually be resumed.

In an Emergency

If you experience severe chest pain, back pain, or loss of consciousness, medical attention is needed immediately. You should call an ambulance or go to the nearest Emergency Department urgently.

As medical staff may not always be familiar with GA disorders, it is vital that you or someone with you tell them about your condition. You may wish to show them this booklet. Some individuals choose to wear a medical alert bracelet in case of emergency. Alternatively, it may be useful to have an emergency information card kept in your wallet at all times.

It may be useful to talk to your partner, family members, or friends about what to do in case of emergency.

Contact numbers for the Marfan and Aortic Clinic at RPA Hospital

During business hours:
+61 2 9516 3456

After hours, the cardiologist on call can be contacted by calling the RPA switchboard on +61 2 9515 6111.
Do I need to make changes to my lifestyle?

The aim of regular monitoring is to promote a long, healthy, and happy life. Some adjustments may need to be made to your lifestyle.

What exercise and sport can I do?
Exercise should be part of your healthy lifestyle, but should be done with caution. Strenuous activities increase the stress on the aortic wall, and therefore increase the risk of enlarging the weakened or re-tearing the aorta. Mild to moderate exercise is generally considered suitable, however this should be discussed with your physician.

Please see the boxes on page 15 for suggested activities that are suitable and those that are not recommended.

Can I continue working?
Most people can continue with their jobs if blood pressure is well controlled. However, physically demanding jobs that require heavy lifting or strenuous activities may be unsuitable. Your physician will discuss what activities to avoid with work situations.

Activities that increase the pressure on the aorta, or excessively increases the heart rate, raises the risk of rupture or dissection

Suitable activities may include:
✓ Walking
✓ Swimming
✓ Kayaking/Canoeing
✓ Tennis
✓ Golf
✓ Dancing

Please consult with your physician to discuss what would be most suitable for your individual situation

Activities that are not recommended include:
X Those that have sudden acceleration and deceleration, such as parachuting or bungee jumping
X Those that increase the aortic pressure e.g. weight lifting
X Those that significantly increase the heart rate e.g. squash, competitive running
X Body contact sports
Other Frequently Asked Questions

Genetic testing – Should family members be screened?
DNA testing is available and is improving in accuracy, but it is not a perfect test. There are pros and cons to consider with genetic testing. Individual circumstances differ and the possibility of genetic testing of family members and children should be discussed with your physician or a genetic counsellor to determine what could be offerred, and what the test results could mean.

What are the risks with pregnancy?
Pregnancy increases the strain on the heart and aortic wall. The risk of dissection increases as the aortic size increases. The major risk of dissection occurs in the last trimester of pregnancy and in the first three months after delivery. Regular monitoring of the aorta during pregnancy and early in the post-partum period is recommended. It is recommended that women considering pregnancy discuss these issues with their physician prior to conception.

Can I drive?
In NSW, cardiovascular conditions such as an aneurysm is considered a health condition likely to affect driving. This means that drivers are required to notify the Driver Licensing Authority, and your physician will assess your fitness to drive based on medical standards as set out by Austroads and the National Transport Commission (NTC). Drivers of heavy vehicles may have to meet specific requirements. Periodic assessments may be a requirement of holding the license. Most individuals have no difficulty meeting the RTA requirements and simply require regular review as part of their usual medical management.

Any additional questions you have can be written in the My Questions section on page 21

Myths and Misconceptions

Vitamins and diet can protect the aorta
A healthy diet is important for cardiovascular health in everyone, but there is no evidence that dietary supplements or any particular diet can protect the aorta. However, high blood cholesterol levels can have an adverse effect on arteries and it is important that you have your cholesterol levels checked and treated if too high.

Life expectancy is poor
Twenty years ago, the outlook for patients with a GA disorder was more limited. In the current era, with active monitoring, medical, and surgical treatment, the life expectancy for individuals with a GA disorder nears that of the general population. An active, full, and long life can be expected.

I can’t travel overseas
There is no restriction on travel, although visiting very high altitudes requires more caution. Individuals with GA disorders have travelled all over the world. Be sure to let your physician know if you planning to travel, so medication and documents can be organised. If you are going to live overseas, medical contacts for medical check-ups in other countries can be arranged.

Only men get aortic disease
The inheritance of GA disorders affects men and women equally. However, it is true that men seem to develop more significant aortic dilation than do women. The reason for this difference is currently unknown.
Coping with the emotional ups and downs

Adjusting to a medical diagnosis and dealing with the associated issues can be an emotional journey of ups and downs. Different people will be affected in different ways, and react in different ways. Some people may feel sad or anxious, whilst others avoid thinking or talking about their diagnosis. It is important to let your physician know about how you are feeling and coping.

Some common experiences reported include: feeling sad, angry, overwhelmed, anxious, crying frequently, unable to concentrate, not sleeping well or sleeping too much, changes in appetite, feeling helpless, having mood changes, and feeling irritable.

Some ways to manage stress include:

- exercising regularly, even a short walk can be helpful
- maintain social contacts and activities
- have a healthy diet
- develop a good sleep routine
- take up relaxing activities like yoga or meditation
- talking to a professional such as a psychologist, or joining a support group

A list of support information and helpful websites can be found on page 25 of this booklet.

My Thoughts and Feelings

Emotional support is an important part of maintaining your health. It is important to express how you feel to others such your physician, family, or friends. This space can be used to write down your thoughts, feelings, concerns.
Questions to Ask Your Physician

Sometimes questions may come up during appointments with your physician, or between appointments, but are then forgotten. This is a list of questions compiled from our discussion with many people with GA disorders, which may help you to get information about your condition and treatment. This list can be used as a reminder of the questions or as a checklist in appointments.

Understanding the diagnosis
- How am I affected by the GA disorder?
- What areas of my body are affected by this condition?
- What are the causes of this condition?

Tests
- How often will I need to have tests?
- What are the procedures for the tests?
- What will the tests tell us?

Prognosis
- What is the life expectancy with this condition?
- What is the best case scenario?
- What is the worst case scenario?

Treatment information
- What treatment is available for this condition?
- What is the purpose of taking medication or having surgery?
- Will there be side-effects from the medication?
- Will surgery be necessary?
- What are the short-term and long-term options?

Lifestyle
- What is the best way to look after myself?
- What sort of activities can I do, what activities should I avoid?
- Are there other sorts of ways to look after myself? e.g. diet, complementary therapies, etc.

Costs
- What will be the costs throughout my treatment?
- Can I claim costs back from medicare or private health fund?

Information and Support
- Is there more information available about this condition?
- Can I talk to someone about how to cope with the stress?

The next page can be used to write down other questions you may have.

My Questions

This space can be used to write down any other questions you would like to ask your physician
My Treatment Plan and Recommendations

This space can be used for important information about your condition, recommendations, and your treatment plan.

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More information and Resources

There are various information sites on the internet relating to GA disorders, and some are listed below. While useful information and support can be sourced from the internet, be aware that not all information is accurate or reliable, and that the information may not be directly applicable to your individual circumstance. It is best to clarify any questions you have directly with your physician.

Marfan Support and Information in NSW
http://marfan.net.au
Information about support networks around Australia are available on the NSW site

International Organisations
International Federation of Marfan Syndrome Organizations (IFMSO) http://www.marfanworld.org/
http://www.marfan.org
http://www.loeysdietz.org/
http://marfan.ca/
http://www.marfan-association.org.uk/
http://www.aorticdissection.co.uk/

Emotional support
Your physician or GP can refer you to a professional such as a genetic counsellor or psychologist.

Research
There is an increasing amount of research being conducted on genetic aortic disorders. Research has continued to improve knowledge about the underlying mechanisms and about treatment advances. As information becomes available, your physician will inform you.
Glossary of Terms

**Angiography:** an x-ray of the blood vessels

**Aorta:** the largest artery in the body that distributes oxygenated blood from the heart to all other parts of the body

**Aortic aneurysm:** an abnormal enlargement or bulging of the aorta

**Aortic dilation:** enlargement of the aorta

**Aortic dissection:** a tear of the aorta wall that allows blood to leak between the layers

**Aortic valve:** guards the entrance to the aorta from the left ventricle

**Blood pressure:** the pressure exerted on the walls of blood vessels by circulating blood

**Computed Tomography (CT) scan:** an x-ray to produce cross-sectional and 3D pictures of the aorta.

"de novo": spontaneous gene variation

**Echocardiography:** type of ultrasound that shows the heart in detail

**Leaflet:** flaps in the aortic valve that regulate the blood flow

**Magnetic Resonance Imaging (MRI):** used to produce cross-sectional and 3D images of the aorta by using strong magnetic field and radio waves

**Open repair:** surgery where an incision is made into the abdomen to expose the aorta and insert the graft

**Stenting:** a stent graft, consisting of a synthetic fabric tube supported by a metal structure, is used to re-line the diseased part of the aorta and divert blood from the aneurysm

**Valve replacement:** procedure to replace the failing aortic valve with an alternate biological or mechanical valve
This booklet was written and developed by:
Joyce Leng, School of Psychology, University of Sydney
Dr Ilona Jurasova, School of Psychology, University of Sydney
Professor Richmond Jeremey, School of Medicine, University of Sydney, Director, Marfan and Aortic
Disease Clinic, Royal Prince Alfred Hospital, Sydney, Australia
with the aim of providing an information resource for individuals diagnosed with genetic aortic
disorders. This booklet is not intended as a substitute for formal medical care or as an authorita-
tive reference of the mentioned conditions.

This material cannot be reproduced or distributed without written permission. For enquiries,
please contact: Dr Ilona Jurasova on +61 2 9036 5275 or ilona.jurasova@sydney.edu.au
Author conflicts of interests: None
Last updated: May 2013

Genetic Aortic Disorders