



Risky business: Insuring adults with congenital heart disease

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Accurate prognostication in congenital heart disease is vital for purposes of obtaining insurance, yet can be problematic for patients, physicians and insurers. This article discusses the scope of the problem, and describes the process of evaluating life insurance. Mortality data as well as predictors of adverse outcomes for individual congenital heart lesions are reviewed. Practical tips for patients and their physicians are given to aid in successful application for insurance. To expand the possibility of future patients obtaining insurance coverage, the ongoing reporting and constant updating of very long-term survival data in congenital heart disease is emphasized. © 2003 Published by Elsevier Ltd on behalf of The European Society of Cardiology.

Introduction

A 35 year old woman with transposition of the great arteries repaired with a Mustard procedure attends your clinic for annual follow-up. Despite her systemic circulation being supported by the right ventricle, she has remained well over the years without need for further intervention. She is not aware of any exercise limitation, works as a secretary and has had a successful pregnancy without complications. There is no history of arrhythmias, and oxygen saturation on room air is 99%. Her main concern, she tells you, is that her recent life insurance application was refused because of congenital heart disease. She is disturbed because at yearly clinic visits she has always been reassured that she is 'doing very well'. She asks you to write to her insurance company on her behalf.

This case highlights a common scenario faced by adults with congenital heart disease (CHD), their physicians and insurers. To the patient whose application for

insurance is refused on the basis of congenital heart disease, there is the perception of being treated unfairly in addition to the unwelcome reminder of reduced life expectancy. Physicians are frequently asked to provide information to the insurance industry regarding the medical history and current status of their congenital heart patients. Apart from the physician's intuition that a patient 'should do well', distilling patient-specific information into a quantitative prognosis can be challenging. Insurers in turn are faced with the task of evaluating information provided to predict outcome compared to a standard reference population in order to determine eligibility for insurance.

The scope of the problem

Congenital heart disease (CHD) affects 8/1000 live births.¹ Due to the development of techniques for the diagnosis and management of CHD over the last fifty years, expected survival into adulthood has reached 85%.² Estimates from the year 2000 suggest that there are close to 150 000 adults with congenital heart disease in the United Kingdom.² Adding to this number are an estimated 1600 cases with complex or significant congenital heart lesions³ and many more with simple lesions entering the adult age group each year. These figures will continue to increase over time, leading to an ever growing adult patient group seeking the financial security of insurance coverage.

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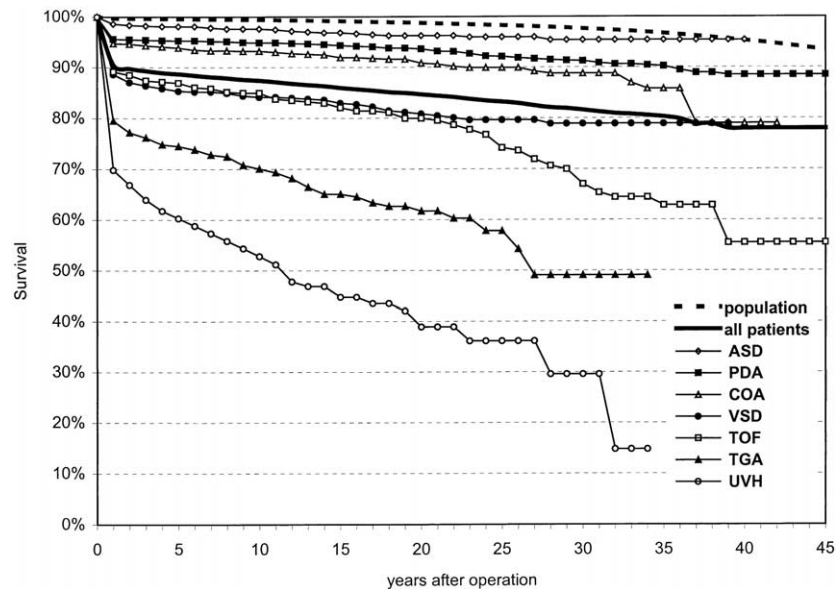


Fig. 1 Survival after pediatric cardiac surgery in a Finnish population of operated patients with congenital heart disease; specific diagnostic categories are compared to a reference population. Used with permission of Dr. H. Sairanen, from Nieminen HP et al. *Circulation* 2001; 104: 573.

The insurance process

Insurance may be sought for a variety of reasons. Individual health insurance serves to cover the cost of medical care, although it usually will not cover costs associated with pre-existing conditions, and often is denied to patients requiring regular cardiac follow-up. Pre-existing health conditions can also preclude obtaining disability insurance to provide income replacement in the event of an inability to continue working. Life insurance pays a benefit upon the death of an individual, and may be desirable for the patient with dependent family members or as part of an application for loans such as mortgages. We will focus primarily on life insurance in the following discussion.

In the evaluation of individuals with chronic illness, the insurance industry attempts to select those patients who are likely to have close to normal longevity out of a pool of similarly affected individuals with worse prognosis in order to avoid issuing excess numbers of policies at high risk of making an early claim. This requires detailed medical information for a given case in order to correctly evaluate and price the risk associated with issuing a policy. Although it is often assumed that an adversarial stance is taken by insurers against patients with chronic illness, companies do want to provide policies to applicants since the insurance industry is competitive and in the risk taking business. So, how do insurers determine how much risk is too much?

For purposes of life insurance, companies consider the expected mortality of a given patient group compared to the observed mortality in a reference population (usually a cohort of insured individuals of the same age) in order to derive a mortality ratio.⁴ For example, patients whose mortality rate is the same as a reference population have a mortality ratio of 100%, while patients with a mortality

rate of 5% over 10 years, compared to a reference population rate of 1% over ten years have a mortality ratio of 500%. For each 100% increase in mortality ratio, the premium paid for insurance is increased by approximately 90%. Patients with a predicted mortality ratio >500% are rarely considered insurable.

Current mortality estimates in adults with congenital heart disease

A number of large scale, long-term studies are available for prognostication of outcomes in some congenital heart lesions. Among the most important of these, is a report from the pediatric cardiac surgical database of 6461 children operated in Finland between the years 1953–1989.⁵ Thanks to a highly accurate and comprehensive national population registry, the authors were able to trace 96% of patients, with a mean follow-up time of 22.3 (range 8.8 to 45.5) years. After exclusion of operative mortality, the late mortality rate over 45 years among congenital heart patients as a group was 16%, compared to 7% for an age-, time- and sex-matched population. Mortality rates for specific lesions with up to five decades of follow-up after surgery were calculated and ranged between 5% for atrial septal defects to 85% for univentricular hearts (Fig. 1).

Broadly speaking, prognosis in congenital heart lesions can be grouped into those lesions with a good outcome (normal or near normal prognosis), those with an intermediate outcome (residual haemodynamic abnormalities and therefore a more guarded prognosis), and those with an uncertain or poor outcome (complex uncorrected anatomy, large variability between individuals with the same lesion, and/or limited data to guide prognosis—Table 1).

Table 1 Prognostic Categories for Adults with Congenital Heart Disease

Good	Intermediate	Uncertain or Poor
Atrial septal defect	Aortic stenosis	Transposition of the Great Arteries
Patent ductus arteriosus	Tetralogy of Fallot	—post arterial switch procedure
Pulmonary stenosis	Transposition of the Great Arteries	Congenitally corrected transposition
Ventricular septal defect	—post Senning/Mustard procedure	Ebstein's anomaly of tricuspid valve
Coarctation of the aorta		Single ventricle physiology

Table 2 summarizes currently published mortality data for common defects from a variety of sources. In addition, where permitted by available data, the best-case scenario mortality rates are shown for low risk patients within each anatomical subgroup. Also shown for each lesion are mortality ratios calculated from the published mortality rates in patients compared to their reference population. For comparison purposes, the last column in Table 2 shows the range of mortality ratios quoted in three insurance underwriting manuals.⁴

Good prognosis lesions

As can be seen from Table 2, among the lesions with a good prognosis (and mortality ratios around 100%) are atrial septal defect (ASD) and patent ductus arteriosus (PDA), especially if repaired at a young age and before the development of pulmonary hypertension.^{5,6} Equally, pulmonary stenosis (PS) has near normal mortality, with early age at operation predicting a better outcome.⁷ Although published studies of early cohorts describe increased mortality rates among adult patients with repaired ventricular septal defects (VSD),^{5,8} in general these patients also have a good prognosis when repaired early, and in the absence of pulmonary hypertension and complete heart block. Coarctation of the aorta (COA) carries a known potential for increased late mortality, primarily due to coronary artery disease.⁹ However, when not associated with a VSD,¹⁰ when operated early (age less than 9) and when postoperative hypertension is absent, long-term prognosis is not far from normal.⁹ Coarctation patients with residual stenosis, or additional lesions such as mitral valve abnormalities, ventricular septal defects, aortic valve disease, as well as those with risk factors for atherosclerosis may have a less favourable prognosis.

Good to excellent prognosis can also be expected in a number of lesions when present in mild form not requiring surgery. These include small restrictive VSD, mild to moderate pulmonary stenosis, bicuspid aortic valve and mild aortic stenosis.

Intermediate prognosis lesions

In contrast to the above lesions with good prognosis, a more guarded prognosis is expected in individuals with congenital aortic stenosis (AS), because potential for late congestive heart failure, infective endocarditis and sudden death¹¹ increases mortality in this group as a whole. Since the spectrum of disease is wide among AS

patients, individual factors such as age, gradient across the valve and post-operative clinical status must be considered in predicting prognosis. Increased mortality compared to the general population is also seen late after repair of tetralogy of Fallot due to propensity for arrhythmias and sudden death. Patients repaired early in life and without a prior palliative procedure¹² have a better prognosis. Favourable outcome is also expected in tetralogy patients without significant pulmonary regurgitation or stenosis (especially those repaired without use of a right ventricular outflow tract patch^{13,14}) and those with preserved right ventricular function. Current adult survivors with transposition of the great arteries (TGA) likewise have greater than average mortality rates. Most patients in the adult age category will have been repaired with a procedure to redirect atrial blood (Mustard or Senning operation). Late survival is impaired in these patients due to systemic ventricular dysfunction¹⁵ and sudden death. Systemic ventricular dysfunction can be detected in at least one-third of patients by 20 years after the Mustard procedure and, along with pulmonary hypertension, increases the risk of late mortality.¹⁶ Positive prognostic factors in TGA include operation in a later surgical era, absence of ventricular septal defect and absence of arrhythmias.¹⁵

Uncertain or poor prognosis lesions

Lack of published data of very long-term follow-up for a number of congenital heart defects makes prognostication difficult. Since the introduction of the Jatene procedure in 1975, the arterial switch has replaced atrial redirection procedures for correction of TGA. Although it is hoped outcome will be improved, insufficient numbers of patients have entered the adult age group to make accurate prognoses. Similarly, accurate predictions of mortality are impaired in uncommon lesions where relatively small numbers of patients have been studied such as congenitally corrected transposition, complex transpositions, Ebstein's anomaly of tricuspid valve and univentricular hearts. Unfortunately, published data as well as experience suggests that outcomes with these lesions will remain substantially poorer than the general population, with estimated mortality ratios over 500%.

Even when long-term data is available for a specific lesion, a host of factors need to be taken into account for accurate prognostication of an individual. Available series generally do not account for the wide spectrum of disease within the same lesion, and may not apply to individuals with multiple lesions. Existing long-term

Table 2 Mortality rates for operated congenital heart lesions compared to a reference population. (Where permitted by available data, mortality in low risk subgroups within each lesion are shown)

Lesion	Reference	Duration of Follow-up (years)	Late Mortality All Patients ^a (%)	Late Mortality Low Risk Subgroup (%)	Mortality in Reference Population (%)	Mortality Ratio All Patients ^b (%)	Mortality Ratio Low Risk Subgroup ^b (%)	Mortality Ratio Insurance Underwriting Manuals ^c (%)
ASD	5	40	5 ^d	N/A	7	71	N/A	100
PDA	5	45	12 ^d	N/A	6	200	N/A	100
PS	10	25	10	6 ^e	6 ^e	167	100	100 up
VSD	7	27	20	5	3	667	167	100–200
COA	8	20	16 ^d	9 ^d	5 ^e	320	180	100–300
AS	11	25	15 ^d	8 ^d	4	375	200	225–400 up
TOF	12	32	14	7	4	350	175	200–400
Senning/Mustard	15	20	24	N/A	5 ^f	480	N/A	Declined
Single Ventricle	4	34	85 ^d	N/A	3 ^e	>2800	N/A	Declined

^aExcludes surgical mortality unless otherwise indicated.

^bCalculated as mortality rate in patients/mortality rate in reference population ×100.

^cRange of mortality ratios published by three insurance companies in underwriting manuals; calculated as mortality rate in patients/mortality rate in reference population of insured individuals ×100⁴

^dIncludes surgical mortality.

^eMortality rate estimated from Kaplan–Meier curve.

^fNo reference population rate given; mortality rate estimated from other studies. N/A: Not available from published data.

outcomes are primarily based on the earliest surgical era (1945–1970). More contemporary cohorts of patients are expected to do better due to improvements in surgical techniques, myocardial protection and peri-operative care. Very little data are available on individuals over the age of 40 or with rarer heart defects. Furthermore, existing published outcomes do not account for future improvements with evolving medical therapies, and for potential interactions with other risk factors that affect outcome, such as smoking.

What patients and their physicians can do

Some practical strategies may be of help to patients seeking insurance. Firstly, declined patients or those offered insurance at high premiums should shop around, as not all companies rate risk the same. For patients with lesions where published data of good outcome are lacking, some companies may be willing to be optimistic and allow coverage with a modest increase in premium. The local adult congenital heart disease patient association may be able to provide advice about insurers with a track record of providing coverage for this growing patient population. Once life insurance contracts are issued, they are not cancellable and remain in force until death.

One important avenue for obtaining health, disability and/or life insurance is through group insurance policies available through employers or professional associations. Group policies do not require an individual evaluation, as they are based on the assumption that the majority of an unselected employee group or association will be healthy. Hence even adult patients with complex CHD may obtain insurance via this route, without need for individual assessment. Patients may want to consider the group insurance benefits likely to be available to them when planning a career or choosing an employer.

Another product that may be available in certain places is the non-renewable term policy which provides short-term coverage ending after a fixed period of time (usually 10 years). These may be useful to some patients until the natural history of their congenital lesion is better understood.

Finally, other factors especially age and smoking status affect insurability. Since mortality in the general population predictably increases with age, whereas the mortality associated with a congenital heart defect may remain the same, mortality ratios of congenital heart patients inevitably decrease with age. Hence those who were uninsurable at age 30, may be able to obtain coverage after age 50. Avoidance of smoking and adoption of good health practices will lower a patient's overall risk, and further increase their chances of insurability. Attention to reducing coronary risk factors to a minimum is more likely to allow a more favourable insurance decision.

Bearing in mind published mortality data for a given lesion (Table 2), physicians making recommendations for an individual patient should present the facts of the case with supporting investigations. When positive prognostic factors are present and negative prognostic factors are absent (Table 3), this should be noted and emphasized to

Table 3 Prognostic factors in adult patients with congenital heart disease

Positive Prognostic Features	Negative Prognostic Features
Biventricular circulation	Early surgical era
Systemic left ventricle	Long standing palliation
Repair at early age	Ventricular dysfunction
Good functional capacity	Arrhythmias
Minor haemodynamic lesions	Pulmonary hypertension
	Complex or multiple lesions

the insurer. Moreover, patients may receive a more favourable response from insurers when followed in a specialized adult congenital clinic where management of CHD and its late complications are routine.

One cannot overemphasize the need for ongoing reporting of very long-term follow-up data for this patient population. Ideally this should be done in a coordinated fashion between centres to yield series large enough to describe outcomes in high and low risk cohorts. Furthermore, it will be of interest to compare contemporary patients repaired after 1980 to those repaired in the early surgical era, as their prognosis is expected to be considerably improved.

Conclusions

Like the general population, adult patients with CHD desire the financial security that comes with insurance. Patients with repaired ASD, PDA, PS and VSD with low mortality ratios should be able to obtain insurance without problem. Patients with intermediate prognosis lesions represent a higher risk group to insurers, but may achieve insurance on the basis of individual consideration, especially in the absence of negative prognostic features. Patients with uncertain or poor prognosis lesions, such as those with complex congenital heart disease, will for the most part be considered uninsurable on an individual basis, although alternative routes via group policies may exist. With newer surgical and catheter techniques, advancing medical therapy and improved risk stratification, overall prognosis and hence insurability will continue to improve for adults with congenital heart disease.

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