Surgical outcomes in congenital heart disease: expectations and realities

William G. Williams*

Hospital for Sick Children and The Toronto General Hospital, University of Toronto, No. 1525, 555 University Avenue, Toronto, Ont., Canada M5G 1X8

Received 8 December 2004; received in revised form 8 February 2005; accepted 11 February 2005; Available online 7 April 2005

Summary

The past 50 years of congenital heart surgery has produced enormous progress. Current results surpass expectations. Yet there are important residual problems in patients growing up after heart surgery for congenital heart disease. Our system of care must evolve to care for these people throughout their lives. The evolution of congenital heart surgery has reached a point in time when we should extend care to patients in under serviced emerging countries. Development of local expertise will be required within those countries that are willing to commit resources to an organized program of caring for people with congenital heart disease. Database technology is an essential tool for ensuring and improving quality of care in every congenital heart centre. Both Registry and Academic databases have much to offer in improving care for future patients. Yet overzealous privacy laws threaten the knowledge base provided by computerized databases. We need to guide our legislators in ensuring that the valuable resource provided by database technology is not lost.

© 2005 Elsevier B.V. All rights reserved.

Keywords: Congenital heart disease; Database; Registry; Academic; Emerging countries adult congenital heart disease

1. Introduction

The first intra-cardiac repair of tetralogy of Fallot (ToF) occurred 50 years ago, with six of the first 10 patients surviving operation [1]. By current standards a 40% operative mortality is unacceptable, but in the context of that era where the only alternatives were a palliative shunt or an unfavourable natural history, Lillehei's accomplishment was monumental.

To day, most patients living with ToF are adults. Their clinical outcomes have been excellent but they are at risk of late complications, especially right heart failure, arrhythmia and a risk of sudden unexpected death. Re-operation during adulthood may be required and is quite beneficial. Progress in heart surgery has permitted primary repair during infancy and may decrease the prevalence of late complications. Early survival close to 100% is now expected.

The evolution of therapy for patients with ToF illustrates both overwhelming success and important failures. These changing expectations and realities of outcomes after congenital heart surgery led Tom Treasure to observe: In the early days (of heart surgery) risk was self-evident…. Expectations are now very high and news of a patient’s death is likely to be met with disbelief and the question ‘What went wrong?’ [2]

The evolution of surgical management of babies with complete transposition of the great arteries (TGA) exemplifies this changing target of expectations and realities. The natural history of neonates with TGA is one of rapid deterioration and early death within days or weeks. Palliative procedures only forestalled early death until the atrial repair devised by Senning (1958) and Mustard (1963) dramatically improved survival from an expectation of death within a few months to an 80% chance of surviving beyond age 20 years.

However the morphologic right ventricle that supports the systemic circulation after an atrial repair may fail and there is an increasing prevalence of late complications. The tremendous expectations from early success are tempered by the reality of late ventricular failure and arrhythmia.

The arterial switch operation for TGA by Jatene (1976) avoids most of the early and late problems of the atrial repair. Early survival rates now close to 99% have greatly exceeded our expectations from the early learning curve of this new operation. Late complications of ventricular failure and arrhythmia are rare. Importantly, the arterial switch operation demonstrated that neonates could tolerate long complex operations very well. It had a profound influence on the early management of other congenital cardiac lesions,
and has fostered the current paradigm of early primary repair or intra-cardiac palliation rather than closed palliative operation and later repair.

These improvements in outcomes for newborns with tetralogy or TGA are two examples of many that illustrate the remarkable progress made in congenital heart surgery during the past 50 years. With this progress we have raised our expectations, and the expectations of our patients and their families to levels that may exceed the realities of living with congenital heart disease (CHD). Our measure of success has for the most part been survival: death being a finite and irrefutable end-point. Lillehei’s initial experience of six survivors among 10 patients provides a simple measure of success that is easily compared to subsequent experience. Conversely by to-day’s standard of early survival for neonates with ToF or TGA approaching 99%, death is a less sensitive measure of clinical expertise, and future studies must rely upon sophisticated statistical analysis including assessment of functional outcomes.

With so much accomplished in the recent past, what are our expectations for the future? And what are the realities that may temper those expectations? I would like to address these questions by touching on four areas of interest, and focus particularly on the importance of database technology in assessing outcomes of congenital heart surgery.

2. **Worldwide availability of congenital cardiac surgery**

At the Monaco meeting of this Association in 2002, there was a memorable symposium on cardio-thoracic surgery in the emerging world. Among the 6 billion people of this world, only 2 billion have access to cardiac surgery.

Care for patients with congenital heart disease is expensive. But amortized over a lifetime it may be very cost-effective. Unfortunately there is no data to compare the cost of congenital heart surgery to other treatment algorithms. Most government funding agencies and all hospital administrators consider only year-end fiscal balance. In competing for resources with patients who have coronary artery disease, patients with congenital heart disease suffer due to the higher acuity of care required in spite of the fact that their long-term benefit may be substantially greater than after coronary artery surgery.

Congenital heart surgery has developed in large urban University-based centres where resources for research and teaching have been available. Now that techniques are established and standardized, is it time it consider moving care for congenital heart disease into the World community? Some emerging areas of the world have far more pressing priorities than rescuing babies with ToF or TGA. However the proportion of countries that are ready for extending care to these children will surely rise.

The paradigm for treating patients with CHD in emerging countries has been to bring them to Western centres. Many cardiac units have provided care for patients from emerging countries and those efforts have showcased on an individual basis what can be accomplished for people with CHD. But to be truly effective, care must move to those under serviced areas by developing local expertise. Dr Carpentier’s model unit in Viet Nam is an excellent case in point [3]. One of our former fellows, the late Dr Ami Cohen developed an outstanding program to foster development of congenital surgery units in several emerging countries [4].

On a global scale it is time to consider a plan to take congenital surgery into the world community and start providing care for the 2/3’s of the planet’s population that has no access.

Tom Pezzella stated that what is required to do so is:

"simply the geographic transfer of 3 basic entities:

1. Ideas and knowledge
2. People
3. Equipment and supplies

What facilitates or hinders this transfer is money and ego” [5].

Our professional organizations could facilitate the development of programs to assess the needs of emerging countries, and a means of providing both prophylactic and active care for children of the world who suffer from cardiac disease.

3. **The emerging adult population of congenital heart disease**

We have inherited the misperception that CHD is a problem of the paediatric population. But we have now reached a point in time when the majority of patients living with CHD are adults, not children [6]. The proportion of adults compared to children living with CHD will continue to increase into the future. This emerging population of adults with CHD require ongoing life-long care as they encounter new problems associated with aging.

The adult with ToF will have pulmonary valve incompetence or stenosis that may lead to right ventricular dysfunction and dilation. By the 3rd or 4th decade of life atrial and ventricular arrhythmia are common and there is an increasing risk of sudden death. These late complications affect obstetrical care, interfere with employment opportunities, and can have profound psycho-social effects. Genetic counselling and support for associated genetic disorders present in about 1/3rd of these people may be required.

Most TGA patients with an atrial repair are now adults. They may develop failure of their morphologic right ventricle and >50% have serious arrhythmia. A small proportion of the arterial switch population has reached adulthood and their late problems are substantially less than after an atrial switch, but they do need ongoing surveillance.

The adult with CHD requires a system of dedicated care by experts familiar with the problems specific to their age group. In our view, this care belongs to an adult hospital and a referral system should be centralized to concentrate care, teaching and research for the complex adult congenital heart patient [6,7].

There should be only a few adult centres. Our figures suggest that each tertiary adult centre should serve

"3. The emerging adult population of congenital heart disease

We have inherited the misperception that CHD is a problem of the paediatric population. But we have now reached a point in time when the majority of patients living with CHD are adults, not children [6]. The proportion of adults compared to children living with CHD will continue to increase into the future. This emerging population of adults with CHD require ongoing life-long care as they encounter new problems associated with aging.

The adult with ToF will have pulmonary valve incompetence or stenosis that may lead to right ventricular dysfunction and dilation. By the 3rd or 4th decade of life atrial and ventricular arrhythmia are common and there is an increasing risk of sudden death. These late complications affect obstetrical care, interfere with employment opportunities, and can have profound psycho-social effects. Genetic counselling and support for associated genetic disorders present in about 1/3rd of these people may be required.

Most TGA patients with an atrial repair are now adults. They may develop failure of their morphologic right ventricle and >50% have serious arrhythmia. A small proportion of the arterial switch population has reached adulthood and their late problems are substantially less than after an atrial switch, but they do need ongoing surveillance.

The adult with CHD requires a system of dedicated care by experts familiar with the problems specific to their age group. In our view, this care belongs to an adult hospital and a referral system should be centralized to concentrate care, teaching and research for the complex adult congenital heart patient [6,7].

There should be only a few adult centres. Our figures suggest that each tertiary adult centre should serve
a minimum general population of 15 million, a population 3-times that required to support a paediatric unit [8].

I recognize that this recommendation to centralize care for the adult CHD patient may appear to be opposite to my views expressed above of taking paediatric cardiac care into the world community. In fact both require centralization and a highly focused commitment to excellence. But because of the relative rarity of the adult with CHD, the critical mass for the adult care unit requires a large referral base.

The emerging population of adults with CHD commonly move to other cities and sometimes to other countries. The network of care for the adult with CHD must therefore accommodate their worldwide migration. Information to facilitate their life-long care anywhere in the world must be easily shared among centres of excellence via Internet.

4. Assessing the outcomes of congenital heart surgery

Dr Lacour-Gayet has stated:

Continuous evaluation of quality of care is becoming a duty of surgical practice [9].

To evaluate quality of care we need accurate data that is collected reliably, systematically and consistently. This data is compiled into a database. A database is nothing more than a structured collection of information [10]. Stark listed the many reasons for a database [10]. The Bristol affair and events in other cities dictate that we have reliable regular surveillance of outcomes data for quality control [11].

Information is the essence of the database. In an ideal world, a database would contain all of the information on all of our patients. To date it has not been possible to obtain and record all of the information on all of our patients. Attempts to collect all data lead to frustration, incomplete data collection, exhausting effort in recording data that is not relevant for some patients, and ultimately failure of the database. However, we are entering an exciting era where it will be possible to have all of the data for all of the patients, not by one large database, but by linking all of the many databases within a cardiac program.

Meanwhile we are limited to the practical alternatives for a database.

Those alternatives are to:

1. Collect some data on all patients
2. Collect some data on some patients
3. Collect all of the data on some of the patients

Option 1 is a Registry database (some data on all patients) and option 3, an Academic database (all of the data on some of the patients) [12]. Option 2, some data on some patients should be avoided and does not warrant a name. It ensures bias and would be a waste of time and effort.

Both a Registry and an Academic database have their own purpose, and each have limitations. There is tremendous interest in Registry databases, including the very successful Society of Thoracic Surgeons, the European Association Cardiac Registry and the Paediatric Cardiac Consortium of University Hospitals database.

To explore similarities and differences in these 2 types of databases, I compare our own Hospital for Sick Children (HSC) surgical division Registry database with the Academic database of the Congenital Heart Surgeon’s Society (CHSS).

Each began in the early 1980’s. In our HSC registry there are currently 12,338 patients, and in the Academic database 3,495 children. In the Registry there are 29 variables per patient and in the Academic database up to 700 variables. The HSC Registry contains 13 megabytes of data whereas there is 140 megabytes in the Academic database.

The maintenance of each database is different. The Registry requires a full-time data manager, a part time programmer and some secretarial assistance, while the Academic database employs three and a half full-time people, including a full-time programmer. In addition we are fortunate to have two consultants for the Academic database in Drs Blackstone and McCrindle. These different requirements are reflected in the annual cost of maintaining each database, $70,000 for the Registry vs. $250,000 for the CHSS Academic database.

The objectives of each database are different. The Registry tracks our clinical activity, provides quality assurance data, and is a tremendous help in initiating clinical reviews.

The objective of the Academic database is to generate new knowledge from sophisticated analysis of specific patient cohorts.

In recent years there is tremendous interest in assessing institutional results. The agreement on an International Nomenclature classification and the widespread participation in centralized, large databases may allow meaningful comparison between institutions or individual surgeons [13].

Can a Registry database make valid comparisons between surgeons and institutions? What additional information can be provided by an Academic database?

4.1. Registry database

We used our HSC database to address the question of the utility of a Registry. We analyzed all patient admissions for surgery from July 1982 to December 31, 2003, during which time; there were 13,392 index operations in children. We explored the utility of two recently devised scoring systems to predict in-hospital mortality; [14] the Jenkins system of Risk Adjusted Congenital Heart Surgery (RACHS) and the Aristotle Basic Score (ABS) [15,16]. We also examined the utility of the British concept of a Benchmark using eight commonly performed operations as an index of overall performance.

The number of patients that are ‘scored’ by each system differs. For the RACHS 85% of our patients are scored, for the ABS 97%, and for the Benchmark only 38%.

Two separate logistic regression models were constructed to evaluate the correlation of RACHS and ABS with the actual in-hospital mortality. Both scoring systems do correlate with in-hospital mortality. The RACHS system was slightly more reliable than the ABS basic score. The correlation between each scoring system and the actual in-hospital mortality is about 70%. Both systems are improved to about 80% by
adding age and weight at index operation, and the era in which the index operation occurred.

We conclude that each scoring system provides a mechanism for risk-stratification that is about 70-80% accurate. Either could be a useful guide for comparing inhospital mortality among surgeons or between institutions. Inclusion of other registry variables, such as age and weight at operation, and the era are important in deriving an equation that most accurately correlates with observed inhospital mortality.

The benchmark scoring system is a simplified alternate method of risk stratifying. However its utility is limited because it incorporates only 38% of the total HSC cohort and is only 55% accurate in predicting in-hospital mortality.

While these scoring systems correlate modestly with inhospital death and provide a useful tool to evaluate differences in mortality between institutions or surgeons, they are only an approximation. There will not be a critical cut-point that determines the difference between good results and bad results. The search for a single numeric value to evaluate performance is probably Utopian. More importantly, these scoring systems do not provide insight into the causes of good and bad results, nor do they lead to changes in management strategy that could improve results.

4.2. Academic databases

The Academic database has the depth of information that can provide insight into causes of success and failure. The information can be mined by sophisticated analytic techniques to uncover 'risk factors' and to illustrate the effects of overcoming those risk factors.

Drs John Kirklin and Eugene Blackstone developed the CHSS database to prospectively study specific neonatal diagnostic inception cohorts of patients. Since 1985, eight groups of infants have been studied. The advantage of pooling data from multiple institutions is obvious as it allows entry of a large cohort of patients over a short time period. The other important advantage of the CHSS academic database is that it includes long-term follow-up, so the analyses are not restricted to in-hospital events.

The Academic database has the ability to study time-related events, either morbidity or mortality. Because death is inevitable, only the length of follow-up before death is variable. Parametric analysis can illustrate the instantaneous risk of death over time (or any other time-related event) by the hazard function, and develop mathematical models that predict probable outcomes for individual patients.

These analyses are complex to perform but simple to comprehend. To illustrate the time-related changing risk of death after operation I have used the hazard function after the 'Perfect' Fontan operation [17] (Fig. 1). The graph shows the instantaneous risk of death on the vertical axis and the time after operation in years on the horizontal axis. The risk of death is high at and immediately after operation, rapidly falls to a low constant risk that is not zero, and begins to rise at about 6 years after operation.

In this portrayal of the instantaneous risk of death after the Fontan operation (or after any cardiac operation), note that there is nothing remarkable about the 30-day or in-hospital time frame that we as surgeons focus upon. Risk is unaffected by an arbitrary 30-day limit or by hospital discharge and continues well after discharge. The factors that affect in-hospital mortality continue to affect outcomes for some time after hospital discharge [18].

The CHSS studies rely upon the data to make conclusions, rather than upon expert opinion. To illustrate the limitations of expert opinion we examined data from the CHSS study of 320 newborns with critical aortic stenosis [19]. We examined the selection criteria for the important decision about whether a neonate would have a bi-ventricular repair or single ventricle palliation; a decision made by the expert physicians caring for these babies. The data determined that 52% of the babies selected for a bi-ventricular repair had a better-predicted survival with a Norwood: their 5-year survival would have improved by a mean of 19%. Among those neonates selected for a Norwood, 17% had a better-predicted survival with a bi-ventricular repair, and a potential increase in 5-year survival of 15%. These data provided a mechanism to decide upon the appropriate care for future babies with critical aortic stenosis.

The CHSS studies use demographic, morphologic, functional, procedural and institutional variables that may affect outcomes. Multivariable analyses of these data identify variables that have a statistically important impact on outcomes. The models constructed from these analyses allow prediction of outcomes and suggest important avenues for fruitful investigation. The CHSS studies are risk-adjusted for each patient rather than risk-stratified by expert opinion into groups of patients of similar risk as in the RACHS and Aristotle scoring systems. In Dr Blackstone's words, the CHSS analyses let the data speak for themselves [20]. These analyses identify factors that impact on outcome and thereby ways and means of improving future results. Paul Sergeant referred to this positive feedback that changes patient management as 'Closing the Loop' [21].

http://www.ctsnet.org/aortic_stenosis_calc/
The academic database has the depth of information to address the question: Are there important differences in institutional outcomes when ‘case-mix’ is included? We have examined this question by using several of the CHSS datasets.

The CHSS study of 710 neonates having a Norwood operation identified eight risk factors for early death after operation, including patient-specific, procedural, and institutional variables [18]. One of the criticisms of these studies is that risk factors identified for the entire cohort may not apply to each individual institution. Indeed that is true, as I will illustrate with birth weight, one of the important variables affecting survival.

After controlling for all other significant risk factors among all 29 institutions entering babies into this study, a baby weighing 2 kg has a significantly lower survival than one weighing 3.5 kg (Fig. 2A). However in the few institutions that had better overall survival with the Norwood operation, birth weight had little effect on survival (Fig. 2B). Conversely, in the high-risk institutions, survival of the smaller baby is significantly lower than a 3.5 kg baby, and worse than the survival in the low risk institutions (Fig. 2C). Therefore, it is true that risk factors identified for the entire cohort of patients may not apply equally to each institution. As these data demonstrate, birth weight is important in most institutions. A few institutions know something important about the management of low birth-weight babies that most others do not. How can we learn from the institutions that appear to have solved a very important clinical problem that confounds most of us?

We attempted to rank institutional performance using multivariable analysis that risk-adjust for each patient (Fig. 3). Most institutions perform within the 70% confidence limits of their predicted survival. Some however are significantly better or significantly worse than predicted after taking into account all known risk factors. Note that institution 18 is ranked number 1 and their results are significantly better than the risk-adjusted survival prediction.

The clinical importance of these variations in institutional performance is illustrated by the outcome of a 2.5 kg baby admitted to either institution 18 or 24. If admitted to institution 18, the predicted 1-year survival is 78%, whereas with all other risk factors controlled, survival is only 55% in institution 24. The net difference in survival after the Norwood operation in these 2 institutions is a 23%. The data concludes that after controlling for all other factors, this important difference in survival is due to institutional performance.

The CHSS series of 408 neonates with pulmonary atresia and intact ventricular septum (PA/IVS) entering any one of 33 institutions also demonstrates substantial differences in institutional survival outcomes [22] (Fig. 4). The risk-adjusted 10-year survival illustrates the variation in institutional performance when all other variables affecting survival are controlled. As in the Norwood operation, most institutions perform within the 70% confidence limits of the predicted risk-adjusted mean value. But there are important outliers that either exceed or fail to meet predicted survival. Note the position of institution 18 that had the very best results with the Norwood operation. Its performance with PA/IVS falls well below expectations.

A competing risk analysis of the PA/IVS data explains some of the variation in institutional survival. The data analysis determined that survival of these babies is highly dependant upon the size of the right ventricle as measured by the Z-score of the tricuspid valve (Fig. 5A). The lines within the graph illustrate the competing outcomes for each
tricuspid valve size. In babies with very small valves only a Fontan path or transplant is possible. Conversely in babies with a normal size tricuspid valve a bi-ventricular repair is both possible and desirable. The line marked 'Death' indicates deaths that occurred before any definitive procedure. Note that mortality is higher in babies with smaller tricuspid valves. In this institution very few children have not undergone a definitive procedure, i.e. are alive with no repair.

The outcomes for this institution illustrate a 'balance approach' to PA/IVS, driven by morphologic data. Babies with the smallest tricuspid valves (Z-score $<-5$) have a 75% chance of having a Fontan operation, and a 20% chance of dying without intervention. At the other end of the spectrum, babies with a normal size tricuspid valve have an 85% chance of having a bi-ventricular repair, and a very low risk of death prior to definitive intervention. The dividing line between protocols of Fontan vs. bi-ventricular repair is a Z-score of about $-2$.

In contrast, an institution with an unbalance approach to PA/IVS that appears to ignore the importance of right ventricular size is shown in Fig. 5B. Bi-ventricular repair was attempted in babies with a very small right heart as indicated by a Z-score $<-2$. They appear to be under-utilizing the Fontan approach; only 10% of babies with the lowest Z-score had a Fontan in contrast to other institutions where 75% had a Fontan. The overall mortality before definitive repair is substantially higher, 80% at a Z-score of $-5$ compared to 20% in the low risk institution. In addition they have a substantial number of children in whom no

---

Fig. 3. Ranking of CHSS institutional performance of the Norwood operation. A vertical bar compares each institutions actual performance to its predicted performance. The code number for each institution is on the horizontal axis. Institutions entering less than 10 children are grouped together. The zero line is the risk-adjusted predicted survival for each CHSS institution. Institutions performing better than predicted are above the zero line. Those performing below predicted survival fall below the line. Note that the 70% confidence limits of each estimate, shown as error bars, overlap the zero line of predicted performance level for most institutions. Some however are significantly better or significantly worse than predicted.

Fig. 4. Institutional ranking of survival for neonates with PA/IVS. The graph format is the same as in Fig. 3. Institution 18 had the best survival for the Norwood operation (Fig. 3), but is significantly below predicted survival for management of PA/IVS.

Fig. 5. (A) The Z-score of the tricuspid valve size is shown along the horizontal axis, ranging from normal size indicated by a Z-score of zero, to 5 SD below normal on the left side of the graph. The vertical axis shows the proportion of children that reach each mutually exclusive end-state within 5 years of their initial admission to hospital. The lines within the graph illustrate the competing outcomes for each tricuspid valve size. This institution has a morphologically driven balanced approach to the management of neonates with PA/IVS. Neonates with a small tricuspid valve are likely to have a Fontan, those with a tricuspid valve close to normal, a bi-ventricular repair. Mortality risk is higher with smaller tricuspid valve size. (2-ventricle, bi-ventricular repair; 1.5-ventricle, 1(1/2)-ventricle repair) (Reproduced with permission from the Journal of Thoracic and Cardiovascular Surgery). (B) The graph shows data from an institution with an unbalance approach to management of PA/IVS that ignores the importance of the size of the right heart. Risk of death is high, few children with small tricuspid valves had a Fontan, and there are a sizable proportion of children who are alive without reaching a definitive state. (Reproduced with permission from the Journal of Thoracic and Cardiovascular Surgery.)
definitive intervention has occurred within 5 years. One assumes a decision about their management has not been made. This institution could improve results by a change in protocol. Their technical performance of surgical operations may be perfectly adequate, but their patients are suffering due to a protocol that ignores the importance of the tricuspid valve size and in their inability to commit some patients to a definitive operation. They appear to lack a morphologically driven protocol.

The important lesson from this illustration of the management of babies with PA/IVS is that careful analysis of the data in the Academic database not only identifies significant variations in performance, but can also identify factors that affect performance and how that performance could be improved. In this example, outcomes are determined by the protocol rather than technical expertise of the surgery.

Institutional excellence in managing one diagnostic group of babies does not necessarily apply equally to the management of other diagnostic groups. There is a substantial difference in survival outcomes among institutions in the CHSS studies of PA/IVS, TGA, interrupted aortic arch and the Norwood operation. No single institution was ranked number 1 in performance in more than one diagnostic group and institutional performance varied considerably.

Clearly we have much to learn from each other. But how can we extend the knowledge gained in one institution to others that are struggling? How can institutions doing poorly learn from others that are doing better? We have not solved this problem in the CHSS and our studies have not had the impact we would wish. Partly this is due to the problems of confidentiality. We do not identify the patients, the institutions or the surgeons, so we do not know whom the best or the worst institution are, nor are we privileged to identify them. We do report to each institution their own results, but they have no way of identifying the outcomes of other institutions. The lack of communication due to confidentiality has lessened the impact of our CHSS studies, and is a disappointment that we have been unable to resolve.

### 4.3. Threats to the future of database development

It is a paradox of the current era that as our ability to generate new knowledge from computerized databases is being realized and as our patients ask for more risk/benefit information, we are increasingly constrained from obtaining important information by changing laws governing the protection of privacy.

In medicine, we learn from our successes and our failures. The evidence is overwhelming that both Registry and Academic databases contribute to our knowledge, identify shortcomings in performance and can be important means in improving outcomes for future patients. But the proven usefulness of databases is threatened by an overzealous concern for confidentiality.

Recent changes in privacy legislation are spreading around the world and will require informed consent for a patient’s data to be included in a database. We support the need for confidentiality. However the selection bias introduced by informed consent may preclude collection of vital information that affects our assessment of patient care.

When the CHSS studies began with the Transposition study in 1985, we did not consider informed consent. No one thought it necessary, and I do not believe it was ever discussed. The study enrolled 978 neonates within 4 years, including 19 babies who died before any intervention. If informed consent had been required, these 19 babies would not have been entered and an important subset of the study would have been lost.

Recent follow-up of the CHSS patients is hampered by “consent issues”. Problems with consent compliance account for 57% of those patients who could not be followed. Most of the parents are willing participants in these longitudinal studies and very appreciative of the interest shown for their child. Refusal to participate is a rare event. Of 1,875 families contacted for follow-up in 2003 only 31 (1.6%) refused to participate.

Tu examined the impact of informed consent on a Registry database [23]. The Canadian Stroke Network Registry links 20 major stroke centres across the country. It is an observational study involving no intervention. Changing legislation that now requires informed consent resulted in a decline in enrolment to only 39% of eligible candidates. Tu identified major selection bias. For example mortality rate was 7% in enrolled patients vs. 22% among those stroke patients who were not enrolled. Importantly, the cost of consent-related issues in this 18-month study was estimated to be $500,000.

Another impressive example of the impact of informed consent occurred in Hamburg [24]. The city of Hamburg maintained a cancer registry for more than 50 years. The data was published annually as a help to physicians and epidemiologists. In the mid 1980’s, new laws were passed requiring informed consent. Participation dropped to 70%. Data were no longer published as the results were considered inaccurate, and the Registry failed.

Unquestionably privacy must be safeguarded. But:

*Public health is threatened by incomplete data more than individual privacy is threatened by disease registries [25].*

Ensuring that the quality of care we provide meets a standard dictates that clinical data must be collected and analyzed. We must be proactive as individuals and as professional Associations to ensure that our colleagues, the public, and the politicians understand the vital importance of data collection.

### 5. Conclusion

During the past 5 decades there have been tremendous improvements in the care of patients with CHD. We have an obligation to maintain the expectations we have set for ourselves and for our patients. Systems of care must be developed for the emerging populations in the underdeveloped world and for the adults who have grown up with CHD. Registry and Academic databases will ensure that our quality of care is good and will help us to realize our expectations for future patients. Much has been
accomplished; the future holds even more promise for improving the care we provide.

Acknowledgements

The privilege of presenting this material is possible only by an extensive group of people that care for our patients. I would like to thank the faculty and staff of the Hospital for Sick Children and the Toronto Congenital Cardiac Clinic for Adults. The members of the CHSS and the staff of the CHSS data centre make these detailed analyses possible.

I would especially like to express my appreciation to M. Gail Williams who has developed, maintained and consistently improved our divisional database. Importantly, her 22 years of outstanding professional service has not conflicted with the nurturing and love of our family.

References

[14] Al-Radi OA, Caldarone CA, McCrindle BW, Jacobs JP, Williams MG, VanArsdell GS, Williams WG. Using the Aristotle basic complexity score and the risk-adjustment congenital heart surgery (RACHS-1) scoring system; is it risk adjustment or not? Accepted for presentation at the annual meeting of the American Association for Thoracic Surgery, San Francisco, California, April 2005.