Cardiol Young 2006; 16 (Suppl. 3): 131–135

ISSN 1047-9511
doi: 10.1017/S1047951106001065

Miscellaneous Topics

Current management of ventricular septal defect

Henry M. Sondheimer, Kristy Rahimi-Alangi

Division of Pediatric Cardiology, The Children's Hospital, University of Colorado School of Medicine, Denver, Colorado, United States of America

Keywords: Congenital heart disease; natural history; Eisenmenger's syndrome

At this time, the current practice for treatment of patients with isolated ventricular septal defect is infrequently studied. With this in mind, it was our intent to assess the current management of ventricular septal defect at a single center, The Children's Hospital, Denver. We reviewed the practice at this institution to determine if there is an evidence base for when or if a patient with an isolated ventricular septal defect requires surgical repair. With approval from the Colorado Multiple Institutional Review Board (protocol # 06-0097), we reviewed the data on patients with isolated ventricular septal defect seen during the calendar years of 2004 and 2005, determining the state of the patients, and the level of intervention through December 31, 2005.

Background

Between 1958 and 1969, the National Institutes of Health of the United States of America funded the Natural History Study of Congenital Heart Defects. During the course of this study, the investigators enrolled 1265 children with ventricular septal defect, cared for at six centers in the United States of America.[1] The centres were in Boston, Buffalo, Baltimore, New York, Rochester, Minnesota, and Denver. Patients with ventricular septal defect and aortic regurgitation were excluded from the study so that it would be possible to determine the frequency of acquired aortic regurgitation in those with a ventricular septal defect. The mean age at enrollment of these patients was seven years, with three-quarters of the patients being under the age of 10. All patients underwent cardiac catheterisation, and were then classified in terms of severity as trivial to mild, with 630 patients falling in this category, moderate to moderately severe, encompassing 355 patients, severe, accounting for 189 of the cohort, with the remaining 98 patients having Eisenmenger's syndrome. The categorization was based on the flow of blood to the lungs, along with pulmonary arterial pressures and vascular resistances.[1] Patients with normal pulmonary arterial pressures were classified as trivial or mild according to the flow of blood to the lungs. Patients with elevated pulmonary pressure, but low pulmonary vascular resistance, were classified as moderate to moderately severe. Patients with elevated pulmonary pressure and elevated pulmonary resistance, who were operable, were placed in the severe category, and those with inoperable pulmonary vascular disease in the Eisenmenger group.

According to the protocol for the study, the treatment was mandated by the classification of each child. Those deemed to have trivial or mild defects were to be managed medically. Those with severe defects were surgically repaired. Those that fell into the moderate or moderately severe categories were managed either surgically or medically on a case by case basis at the discretion of their individual cardiologists. Those that were classified with Eisenmenger's syndrome were considered inoperable, and were managed medically. Of the 355 children found to have moderate or moderately severe defects, 230 were initially managed medically, giving a total of 860 patients who followed medical management, not counting those with Eisenmenger's syndrome. Within this group 245 patients eventually went on to have surgery. Of those patients, 149 had initially had trivial to mild defects, accounting for 23.7 percent of the original group, and 96 had initially had moderate or moderately severe

Correspondence to: Henry M. Sondheimer MD, Division of Pediatric Cardiology, B100, The Children's Hospital, 1056 East 19th Avenue, Denver, CO 80218-1008, United States of America. Tel: +303 861 6820; Fax: +303 837 2595; E-mail: sondheimer.henry@tchden.org

defects, accounting for 41.7 percent of this initial group undergoing medical management. The likelihood of surgical repair being the final outcome among those first managed medically was correlated with right ventricular hypertrophy, cardiomegaly, ratio of flow of blood to the lungs relative to systemic flow greater than or equal to 1.5, and the presence of congestive heart failure at entry into the study.[2]

In 1985, the Second Natural History Study of Congenital Heart Defects was begun to generate long-term follow-up data on the patients from the first study, and potentially to shed new light on clinical management of this population. Between 1985 and 1990, 976 of the original cohort of 1265 patients were successfully contacted. A questionnaire was completed by 255 patients, while 570 of the surviving 1099 patients participated fully. Those participating fully included 318 of those initially having trivial or mild defects, 172 of those with moderate or moderately severe defects, 55 with severe defects, and 22 of the group with Eisenmenger's syndrome. Those that chose to participate fully had a median age of 28.9 years, and follow-up evaluation occurred at a median of 22.1 years after their participation in the first study. These 570 patients underwent a full clinical examination, and at least one of a resting electrocardiogram, chest X-ray, echocardiogram with Doppler readings, exercise testing, or Holter monitoring.[2]

Following the first study, morbid events had occurred 215 times over a total of 17,481 patient-years of follow-up. The events reported were bacterial endocarditis, congestive heart failure, brain abscess, syncope, angina, myocardial infarction, stroke, and implantation of a pacemaker.[2] Such events had occurred 1.7 times more frequently in those managed surgically than in those having medical management.

Among the patients that were managed medically, only three developed pulmonary hypertension, defined as a tricuspid regurgitation detected by Doppler and greater than 3 metres per second. Of those patients, two had initially had trivial or mild defects, accounting for 0.6 percent of this group, while the other patient had a moderate or moderately severe defect, again accounting for 0.6 percent of the original group. Only one of the patients who had been classified as having a severe lesion, 1.8 percent of the group, presented with pulmonary hypertension at the time of the second study when using the same definition depending on tricuspid regurgitation using Doppler interrogation of greater than 3 metres per second.[2]

As emphasized above, co-existing aortic regurgitation at the time of entry had been a criterion for exclusion from the first study. At the time of the second study undertaken from 1985 through 1990, only 12 of the 570 patients (2.1 percent) had developed aortic regurgitation. Of these twelve, three had been managed medically and nine had undergone surgery. Of the nine with surgery, eight developed aortic regurgitation after closure of the ventricular septal defect, and only one before surgery. It was twice as likely, therefore, that patients would develop aortic regurgitation after surgical closure of the defect when assessed over the course of the two studies. None of these twelve patients went on to require replacement of the aortic valve.[2]

Congestive heart failure occurred in 126 of the 570 patients reviewed in the second study. Of those, 20 had Eisenmenger's syndrome, leaving 106 patients that were either medically managed or surgically repaired. Medically managed patients accounted for 34 of these patients while the other 72 patients had undergone surgical repair.[2] It is unclear how many of the patients undergoing surgical closure had congestive heart failure prior to surgery, and how many developed congestive heart failure after surgical repair. Failure requiring pharmacologic treatment developed in 22 patients after enrollment in the first study. Out of 322 medically managed patients, 12 (3.7 percent) required one or more cardiac medications for management, while 10 out of 225 (4.4 percent) surgically managed patients required cardiac medication.[2] These numbers exclude those with Eisenmenger's syndrome.

Of the 567 patients for whom data was available, 90.7 percent had a functional classification within the system of the New York Heart Association of class 1 during the later study. When patients with Eisenmenger's syndrome are excluded, the percentage of patients in the first class increases to 94 percent.[2]

Methods

In 2006, we performed a retrospective chart review at The Children's Hospital of Denver of the 19,215 echocardiograms performed in the calendar years of 2004 and 2005. The echocardiographic records for those years were queried for ventricular septal defect. Patients with major cardiac conditions, such as Tetralogy of Fallot, Shone's syndrome, atrioventricular septal defect, transposition, double outlet right ventricle, pulmonary atresia, and tricuspid atresia, which often include a ventricular septal defect, were eliminated from further review. Those with ventricular septal defect and cardiac conditions, such as an atrial septal defect in the oval fossa, patency of the oval foramen or arterial duct, and coarctation of the aorta were retained. In addition, any patient who had had surgical repair of a ventricular septal defect prior to 2004 was eliminated. In the end, we identified 540 subjects with isolated ventricular septal defect. Of those 540 patients, 79 had surgical closure of their ventricular septal defects during 2004 and 2005, while the other 461 were being medically

managed as of December 31, 2005. During the two years of the study, no child had closure of the defect by interventional catheterisation.

Our goal was to try to clarify the indication for surgery in each of the 79 patients undergoing operative closure, and establish what distinguished them from those who were judged not to need surgery. Management had been determined by the cardiologist of each child.

Size and location of the defects

The majority of the defects, 339 of 540, (63 percent) were in the muscular septum, but only 15 of the patients with such muscular defects (4 percent) required surgery. Of the 196 patients with perimembranous defects, in contrast, 61 (31 percent) were deemed to require surgery, and three of the five children with doubly committed outlet defects needed surgery. All patients with multiple defects were classified as perimembranous, muscular, or outlet according to the location of the single largest defect (Fig. 1).

The great majority of the muscular defects, specifically 309 of 339 (91 percent) were deemed to be small, having a diameter of less than 3 millimetres. None of the children with these small muscular defects needed surgery. There were 15 children with muscular defects of moderate size, from 3 to 5 millimetres in diameter, and 3 of them needed surgery. Of the 15 children with large muscular defects, greater than 5 millimetres in diameter, 12 needed surgery. Of the perimembranous defects, the majority, 119 of 196, or 61 percent, were again small, and only one patient with such a small defect required surgery. There were 28 patients with perimembranous defects that were moderate or large, and 16 of these required surgery. In all, 49 patients were found to have large perimembranous defects, and 44 required surgery. None of the doubly committed outlet defects were small, 2 patients having moderately large defects, with neither requiring surgery, whereas surgery was performed for all three children with large outlet defects (Fig. 1).

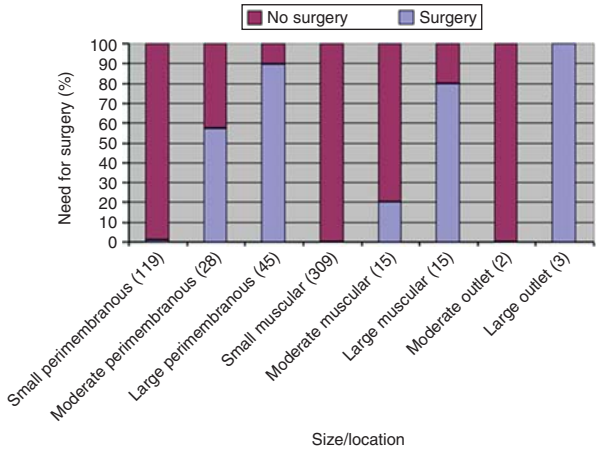

Figure 1.
The chart shows the proportions of defect requiring surgical closure according to their size and morphology.

Surgical management

Surgical repair was performed in 79 of the patients seen at The Children's Hospital of Denver with the diagnosis of isolated ventricular septal defect during 2004 and 2005. In all 79 patients, a Gore-Tex patch was used to close the defect or defects.

Their median age was 134 days, and the average was 265 days. Of the children, 50 had surgery before the age of six months, and 66 (84 percent) before one year of age. The ages at the time of surgery ranged from 4 days to 8 years. The indications for surgical repair are shown in Figure 2. As can be seen, they included aortic regurgitation in 4, double chambered right ventricle in 1, congestive heart failure in 30, pulmonary hypertension in 62, associated aortic coarctation in 2, and failure to thrive in 3. Congestive heart failure was defined by the need for at least one daily therapeutic medication. Pulmonary hypertension was defined either by Doppler echocardiographic findings of tricuspid regurgitation of greater than 3 metres per second, or shunting across the ventricular septal defect of less than 3 metres per second. Failure to thrive was defined by being less than the third percentile in weight for age.

Pulmonary hypertension

Of the 79 patients requiring surgical repair, pulmonary hypertension was the indication in 36. Moderate pulmonary hypertension, with flow across the ventricular septal defect as judged by Doppler

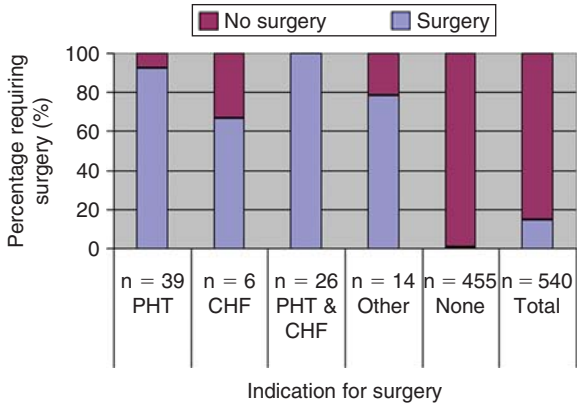

Figure 2.
The chart shows the numbers of patients undergoing surgery in the presence of potential criterions for surgical closure. Abbreviations: PHT: pulmonary hypertension; CHF: congestive heart failure; Other: see text for indications.

https://doi.org/10.1017/S1047951106001065 Published online by Cambridge University Press

from 2 to 3 metres per second, or tricuspid regurgitation of 3 to 4 metres per second, was found in 15 of these patients, while severe pulmonary hypertension, with shunting of less than 2 metres per second, tricuspid regurgitation of greater than 4 metres per second, was present in 21.

Congestive heart failure

Children with congestive heart failure were judged to be controlled or uncontrolled by their medication depending on their growth curves. Congestive heart failure was judged to be controlled with medication if the child maintained a growth curve above the third percentile of weight for age, but was deemed to be uncontrolled if the child was below the third percentile of weight for age. Of the patients requiring surgical closure, 30 had congestive heart failure. Of these 30, 26 also had at least moderate pulmonary hypertension. Of the four patients who did not have signs of pulmonary hypertension, but were being treated with medications for congestive heart failure prior to their surgery, 2 could not be controlled, while the failure was controlled by the medications in the other two.

Pulmonary hypertension together with congestive heart failure

Both pulmonary hypertension and congestive heart failure were indications for surgery in 26 patients. Amongst these 26 patients, 3 had controlled congestive heart failure and moderate pulmonary hypertension, 7 had controlled congestive heart failure and severe pulmonary hypertension, 7 had uncontrolled congestive heart failure and moderate pulmonary hypertension, while the remaining 9 had uncontrolled congestive heart failure and severe pulmonary hypertension.

Other patients

In 13 patients, there were no signs of either congestive heart failure or pulmonary hypertension, but they were still put forward for surgical repair due to other medical conditions. These included aortic regurgitation in 5, double chambered right ventricle in 1, and failure to thrive in 3. Of the remaining patients, 2 underwent repair in the first six weeks of life due to the co-existence of severe aortic coarctation. The final two children did not meet any of the chosen criterions for surgical closure. Of these two, one was at the fifth percentile of weight-for-age. The second patient, albeit without any of the above indications for surgery, was eight years old, and had a concomitant resection of a subaortic fibrous shelf along with closure of his ventricular septal defect.

Medical management

Of the 540 patients diagnosed as having isolated ventricular defect during 2004 and 2005, 461 patients were managed without surgery. Medical management was defined as a patient who was followed in clinic by a cardiologist without any intervention to repair the ventricular septal defect.

Population

The age for the group as at December 31, 2005, ranged from 1 day to 19 years. The mean age was 3.6 years, and the median age was 1.8 years. Overall, 257 (55.5 percent) were two years of age or younger at the cut off date for collection of data, while 90 (19.4 percent) were six years of age or over (see Fig. 1).

Potential indications for surgery

Of the 461 patients being managed medically, 8 also met our criterions for surgical repair. Of these, 3 had moderate pulmonary hypertension, 2 had congestive heart failure that required medication, and three had aortic regurgitation. The defects in the remaining 455 patients were either closing, or had closed, were pressure restrictive, or no longer produced signs of congestive heart failure or pulmonary hypertension (Fig. 2).

Pulmonary hypertension

All 3 patients with pulmonary hypertension undergoing medical management were under the age of 1 year, with 2 of the 3 undergoing surgical closure early in 2006. The third patient showed signs of closing the ventricular septal defect, and the pulmonary hypertension was diminishing. That patient continues to be monitored to see if the defect will close sufficiently to remove the need for surgery, or at least become small enough to justify occlusion by a device inserted in the catheterisation laboratory.

Congestive heart failure

The two patients with congestive heart failure that required medication both satisfied the criterions for control, since they maintained a growth curve above the third percentile of weight-for-age. Their ages were 94 days and 139 days, respectively. Both patients were being followed monthly or bimonthly at the end of collection of data.

Other patients

In this population, 3 patients were found to have aortic regurgitation, 2 of these being aged from 5 to 10 years, and the other in the group aged from 15 to

19 years. The majority, 453 out of 461 (98.3 percent) of patients managed medically did not exhibit any of the potential indications for surgery. Review of the charts and echocardiograms of these patients at the end of the study indicated that the defects were closing in 131 cases, had closed in 81, had become pressure restrictive in 64, or in the remaining 177, the child did not exhibit any signs of congestive heart failure or pulmonary hypertension.

Conclusions

We conclude that the risk of medical management of children with ventricular septal defect who have neither severe pulmonary hypertension nor congestive heart failure is extremely low. Of our large cohort of children with ventricular septal defect seen in 2004 and 2005, only one-sixth, 79 from 540, required surgical correction. If our findings are representative, then the large majority can be managed expectantly with a high likelihood of closure, a low risk of pulmonary hypertension, and a low likelihood of the development of aortic regurgitation. As new options for closure of ventricular septal defects become more widely available, specifically device closure in the catheterisation laboratory, it is important to remember that very few patients who require closure are able to have their intervention delayed into a period when closure using the device would be applicable. Most of those with severe congestive heart failure, and/or significant pulmonary hypertension, will need surgical closure by the age of six to nine months. In our group, the indications for surgery among those having their defects closed beyond the age of one year include aortic regurgitation, double chambered right ventricle, and subaortic stenosis. These concomitant conditions will frequently preclude consideration for non-surgical closure of the defects.

References

1. Weidman WH, Blount Jr, SG DuShane JW, Gersony WM, Hayes CJ, Nadas AS. Clinical course in ventricular septal defect. Circulation 1977; 56 (Supp 1): I56–I69.
2. Kidd L, Driscoll DJ, Gersony WM, et al. Second Natural History Study of Congenital Heart Defects: Results of treatment of patients with ventricular septal defects. Baltimore, Maryland, Circulation, 1993; 87 (Supp 1): I38–I51.

https://doi.org/10.1017/S1047951106001065 Published online by Cambridge University Press