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Cardiac Operations for North American Children with Rheumatic Diseases: 1985–2005

Cory Stingl,

Division of Pediatric Rheumatology, Department of Pediatrics, University of Minnesota Amplatz Children's Hospital, 420 Delaware St. SE, MMC 296, Minneapolis, MN 55455, USA

James H. Moller, and

Division of Pediatric Cardiology, Department of Pediatrics, University of Minnesota Amplatz Children's Hospital, 420 Delaware St. SE, MMC 296, Minneapolis, MN 55455, USA

Pediatric Cardiac Care Consortium, University of Minnesota, Minneapolis, MN, USA

Bryce A. Binstadt

Division of Pediatric Rheumatology, Department of Pediatrics, University of Minnesota Amplatz Children's Hospital, 420 Delaware St. SE, MMC 296, Minneapolis, MN 55455, USA

Bryce A. Binstadt: binstadt@umn.edu

Abstract

Certain pediatric rheumatic diseases are known to affect the heart, sometimes requiring surgical intervention. The Pediatric Cardiac Care Consortium database was used to characterize cardiac surgical intervention among children with rheumatic diseases from 1985 to 2005. From this large database, the records for patients younger than 21 years who underwent cardiac surgery for any rheumatic disorder were extracted. The data collected included the type of procedure performed, the age at the time of the procedure, and the year the procedure was performed. The 261 pediatric patients identified underwent 361 cardiac surgical procedures for complications of rheumatic heart disease (RHD; 160 patients), neonatal lupus (NLE; 53 patients), Kawasaki disease (KD; 28 patients), systemic lupus erythematosus (SLE; 13 patients), and juvenile rheumatoid arthritis (JRA; 7 patients). Multiple procedures were performed for 23% of the patients. The most common procedures included pacemaker implantations among infants with NLE, coronary artery bypass grafts for KD primarily in 5- to 15-year-olds, and cardiac valve operations among adolescents with RHD, SLE, and JRA. Six perioperative deaths occurred. The proportion of annual pediatric cardiac surgical volume attributable to rheumatic diseases did not change during the period studied. Despite advances in their medical care, children with rheumatic diseases continue to sustain measurable morbidity and mortality due to the cardiovascular manifestations of their disease.

Keywords

Arthritis; Kawasaki; Lupus; Rheumatic; Surgery

The associations between specific pediatric rheumatic diseases and their most common cardiac complications are well recognized. Rheumatic fever is associated with valvular carditis (rheumatic heart disease [RHD]), systemic lupus erythematosus (SLE) and juvenile rheumatoid arthritis (JRA) with pericarditis, Kawasaki disease (KD) with coronary artery aneurysms, and

neonatal lupus erythematosus (NLE) with congenital atrioventricular block, among others. Despite these well-known associations, the absolute number of children with cardiac complications of rheumatic diseases, particularly complications severe enough to warrant surgical intervention, is low. As a result, data characterizing these most severely affected children are sparse.

Several recent reports describe operative intervention for RHD in children of North America [9,19] and the developing world [3], but the number of patients is limited because these are single- or dual-institution reports. Excellent national surveys examining trends in coronary artery bypass grafting for KD in Japan have been reported [24,25,34], but similar data for North America are lacking. For SLE, reports describe echocardiographic abnormalities [1,18,20, 32], but information regarding operative intervention for children with SLE is very limited [11].

Thus, except for a large registry report describing 113 infants with congenital heart block (primarily due to NLE) [6], few multicenter studies exist regarding operative intervention for pediatric rheumatic diseases in North America. We therefore used the multi-institutional Pediatric Cardiac Care Consortium (PCCC) database to characterize cardiac surgery among children with rheumatic disorders in North America over a 21-year span.

Patients and Methods

Description of Database

The PCCC is a collaborative network of pediatric cardiac surgical centers in North America that collects information on procedures performed for both congenital and acquired heart disease in children [29,30]. Each patient has a unique identifier to allow longitudinal tracking within the database. We searched the database for records containing diagnostic codes for arthritis (JRA), KD, neonatal lupus, rheumatic fever (acute or chronic), sarcoidosis, scleroderma, SLE, and vasculitis, and for the terms “antiphospholipid,” “dermatomyositis,” and “spondyloarthropathy.” Cases of surgical intervention were found for RHD, NLE, KD, SLE, and JRA, but for none of the other diagnoses. This study was approved by the Institutional Review Board of the University of Minnesota.

Exclusions

This study included only pediatric patients younger than 21 years at the time of their first procedure. Procedures performed at non-PCCC sites before a patient’s admission to a PCCC site were excluded due to insufficient data except for age at the time of first device implantation for patients with NLE. We also excluded the following types of procedures from the analysis: catheterizations, diagnostic procedures, atrial appendage surgeries occurring concomitantly with another surgical procedure, and extracorporeal membrane oxygenation.

Analysis

The surgical procedures included for further analysis were grouped into categories based on type of procedure. Procedures performed on multiple cardiac valves during one operation were counted separately. Multiple procedures performed on the same anatomic structure during one operation (e.g., valve annuloplasty and valvuloplasty on the same valve) were considered one procedure. Autografts of a healthy pulmonary valve into the aortic position to replace a damaged aortic valve (Ross procedure) were counted only in the aortic valve category. Coronary artery bypass procedures were counted as one procedure, and the number of vessels bypassed was recorded.

In-hospital mortalities were reported using available PCCC data. Follow-up data after final discharge of a patient from a PCCC center were not available unless that patient had been readmitted to a PCCC center for another operation.

The annual number of cardiac surgical procedures for pediatric rheumatic diseases was normalized to the annual number of all cardiac surgical procedures among patients 21 years old or younger at the time of their initial procedure. This figure is reported as the number of procedures for the particular disease per 100,000 PCCC surgical procedures.

Results

Between 1985 and 2005, the annual number of PCCC institutions admitting patients younger than 21 years ranged from 14 to 41, and the total number of cardiac operative admissions for patients younger than 21 years was 94,094 (Fig. 1). From this large pool, we identified 261 patients who had undergone cardiac surgery for complications of RHD, NLE, KD, SLE, or JRA. These patients with pediatric rheumatic diseases underwent a total of 361 cardiac surgical procedures, and 61 patients (23%) had more than one procedure, resulting in an average of 1.4 procedures per patient. Six perioperative deaths occurred (Table 1). Most (91%) of the patients with RHD were from North America (United States, Canada, Mexico, Caribbean islands). No patients with NLE, KD, SLE, or JRA were from outside North America.

Among the patients with pediatric rheumatic diseases, RHD accounted for the majority of the cardiac surgical volume (Table 1; Fig. 2). Valve repairs and replacements accounted for the vast majority of the procedures performed for RHD (Fig. 2). Table 2 describes which valves were involved in each operation. Left-sided valves were involved in all but one case, which required an isolated tricuspid repair. Right-sided valves were included in 10.9% of the procedures, with only one case involving a pulmonary valve (excluding Ross procedures) (Table 2).

Among all the valve operations for RHD, approximately two-thirds were valve replacements and one-third were valve repairs. The 93 aortic valve procedures comprised 16 Ross procedures, 61 other valve replacements, and 14 valve repairs. The database entries for two aortic operations did not describe the exact procedure performed. Patients undergoing surgery for RHD were typically 5 years of age or older (Fig. 3).

As expected among patients with NLE, congenital heart block was the most common diagnosis requiring surgical intervention, with 82% of the procedures performed for pacemaker implantation or reimplantation and 5% for device revision (Fig. 2). Half of the 50 NLE patients who had their first pacemaker implanted at PCCC centers underwent one or more pacemaker replacements at a PCCC center. This explains the finding that patients with NLE had the highest percentage of multiple procedures among the conditions studied (Table 1).

Initial pacemaker implantations were performed primarily for neonates, whereas pacemaker replacements were distributed across the pediatric age range (Fig. 3). Six patients with NLE had cardiac transplants. Interestingly, 14 patients with NLE also had structural congenital heart disease including atrial septal defect, patent ductus arteriosus, ventricular septal defect, anomalous pulmonary venous drainage, pulmonary atresia, situs ambiguous, and anomalous origin of the left coronary artery from the pulmonary trunk.

Kawasaki disease was the third most common pediatric rheumatic disorder leading to cardiac surgical intervention (Table 1). As Fig. 2 shows, 19 (61%) of the procedures were coronary artery bypasses. The distribution of bypass procedures included 58% single-vessel, 32% double-vessel, and 10% triple-vessel operations. Seven procedures (23%) were performed for cardiac valve dysfunction (Fig. 2). None of the KD patients with valve operations had coronary

artery bypass surgery. The patients undergoing cardiac surgery for KD tended to be 5 years of age or older (Fig. 3).

We identified 13 patients with pediatric SLE who underwent cardiac surgical procedures (Table 1). Among the 15 procedures performed, eight (53%) were valve repairs or replacements, and four (27%) were for pericardial disease (Fig. 2). No heart transplantations occurred in this group. The SLE patients requiring cardiac surgery tended to be adolescents, consistent with the typical age at onset of pediatric SLE (Fig. 3).

Seven patients with JRA underwent cardiac surgical intervention (Table 1) including valve repairs, pericardial procedures, and device implantations (Fig. 2). These patients were primarily older (Fig. 3).

The proportion of annual cardiac surgical volume attributable to pediatric rheumatic diseases varied substantially (range, 47–667 per 100,000 pediatric surgical procedures), with no obvious trends in this proportion between the years 1985 and 2005 (Fig. 4).

Discussion

The associations between certain pediatric rheumatic conditions and their cardiovascular sequelae are well known. Because these rheumatic conditions occur infrequently in North America, the absolute number of patients who experience severe cardiac morbidity and mortality is low. We have taken advantage of a large North American multicenter collaborative network to characterize cardiac surgical intervention among patients with pediatric rheumatic diseases.

Rheumatic Heart Disease

Worldwide, RHD remains the most common etiology of acquired heart disease in children [7,8]. Higher rates of operative intervention for RHD are reported in the developing world than in North America. For instance, a report from one hospital in Lebanon described 91 patients with rheumatic fever hospitalized between 1980 and 1995. Of these patients, 93% had RHD, with 28 patients (33.7%) requiring operative intervention [3].

In North America, the incidence of rheumatic fever has declined substantially over recent decades, leading to less cardiac surgical intervention. In the year 2000, pediatric rheumatic fever accounted for 503 hospitalizations in the United States, for a rate of 14.8 cases per 100,000 hospitalized children [28]. Among these hospitalized patients, 30.4% had cardiac involvement, and 2% of the hospitalizations (6 cases) involved cardiac surgical repair [28]. A retrospective analysis of 596 patients with rheumatic fever seen at a single institution in the United States between 1985 and 2003 showed that 26 of 366 patients with RHD (7.1%) required operative intervention [19]. Similarly, a study of 98 patients with rheumatic fever from two pediatric hospitals in Montreal between 1979 and 2005 reported 6 valve replacements among 71 patients with carditis (6.1%) [9].

The current study included 160 pediatric patients primarily from North America who underwent 200 surgical procedures for RHD over 21 years. The most frequent types of procedures and the age distribution of the patients are similar to those in prior reports [3,9, 19].

The large size of the PCCC database also allowed us to detect procedures performed rarely for RHD, including cardiac transplantation required by two patients. Thus, RHD continues to result in measurable cardiac morbidity and mortality among children, even in developed countries. This highlights the need for continued early recognition and treatment of streptococcal

pharyngitis, secondary prevention for patients with rheumatic fever [7,15], and improved detection of subclinical rheumatic heart disease [27].

Neonatal Lupus

Congenital heart block and other manifestations of neonatal lupus result from the transplacental passage of anti-SSA/Ro and/or anti-SSB/La antibodies from the mother to the developing fetus [22]. The majority of patients with congenital heart block require pacemaker implantation, but the percentage of patients requiring pacing varies in different studies. A Finnish study reported 53% of patients required pacing [14]. A North American registry study reported 63% of patients requiring pacing, most often in the first days of life [6], and a Swiss study reported that 81% of patients required pacemakers [2]. A Canadian study reported that by the age of 20 years, only 11% of patients with neonatally diagnosed congenital heart block were not paced [23].

By providing longitudinal analysis of 55 patients with NLE, our study offers additional insight regarding the long-term surgical course of these patients, including the need for pacemaker revision and even cardiac transplantation. These data provide additional motivation for efforts aimed at understanding the pathogenesis of congenital heart block and the development of effective preventive strategies [22].

Additionally, we noted the presence of structural congenital heart defects in 26.4% of patients with NLE. Other studies have reported this as well, with rates ranging from 14 (12.4%) of 113 infants to 38 (42%) of 90 infants with NLE [6,14]. Similar to our data, the most common abnormalities included atrial septal defect and patent ductus arteriosus. These defects are not thought to be directly related pathogenically to the presence of anti-SSA or anti-SSB antibodies [6]. A proposed mechanism is congenital heart block that causes asynchronous contraction of the atria and ventricles and interferes with proper closure of the foramen ovale, leading to atrial septal defect [14]. These observations suggest the need for a more focused investigation to determine how autoantibodies influence cardiac development.

Kawasaki Disease

Kawasaki disease is the leading cause of acquired heart disease among children in Japan and North America [5]. The incidence of KD in Japan (108/100,000 in 1996) is several times higher than in the United States (17.1/ 100,000 in 2000) [21,35]. Several excellent studies from Japan have reported the outcomes for large numbers of children in whom coronary artery bypass grafts have been performed for sequelae of KD [24,34]. Kitamura et al. [25] recently reported a 20-year survival rate of 98.4% among 110 Japanese patients with KD in whom bypass was performed. Similar data for North American patients are sparse and confined largely to case reports or small case series [17]. Our data also highlight the fact that patients with KD may have not only involvement of the coronary arteries, leading to the need for bypass, but also valvular involvement severe enough to warrant valve repair or replacement. The age distribution of the KD surgical cohort we report is similar to that of prior reports [17,34].

Systemic Lupus Erythematosus

Most reports of cardiac involvement in SLE describe echocardiographic findings in adult patients [10,31,33]. Reports of echocardiographically defined cardiac involvement in pediatric SLE also exist. A study of 31 pediatric patients reported 7 with pericarditis, 5 with myo/pericarditis, and 1 with myocarditis [32]. Additional studies have reported valvular regurgitation in 5 of 14 and 8 of 40 pediatric SLE patients [1,18]. A recent description of 256 pediatric SLE patients reported 39% with pericarditis, 6% with myocarditis, and 1% with endocarditis [20]. Thus, patients with pediatric SLE commonly experienced cardiac complications. Reports of surgical intervention for cardiac complications of SLE, however, are infrequent. One report describes surgical intervention for nine adults with SLE including

six coronary artery bypass grafts, two valve replacements, and one combined cardiac and renal transplantation [26]. In pediatric SLE, a single case report of a mitral valve replacement for a 17-year-old with lupus has been published [11].

Our study provides additional insight regarding the requirement for cardiac surgery among pediatric patients with SLE. Although pericarditis is observed more commonly than endocarditis in both pediatric and adult SLE [13,20], we found that valvular disease accounted for the majority of cardiac surgical procedures among pediatric patients with SLE. This presumably reflects the fact that pericarditis is more easily managed pharmacologically or via pericardiocentesis. Our report should serve as a reminder that cardiac valve involvement, although uncommon among children with SLE, can progress to the need for surgical intervention.

Juvenile Rheumatoid Arthritis

Juvenile rheumatoid arthritis (JRA) and juvenile idiopathic arthritis (JIA) can lead to pericarditis, myocarditis, or endocarditis. Cardiac involvement is a concern primarily among patients with systemic juvenile arthritis, although patients with other JRA subtypes and cardiac complications have been reported [4,12,16]. The database we used did not contain information regarding the subtype of JRA each patient had. Similar to other reports, however, pericardial and valve procedures accounted for the majority of procedures among patients with JRA.

Study Limitations

Because this study was retrospective and relied on an existing database, certain data could not be obtained, leading to some limitations of the study. For instance, longterm outcome data are not included because the database contains only interventional records from its participating centers. Therefore, patients whose subsequent visits are noninterventional or occur at other sites are not available for analysis. Importantly, the PCCC database also does not include information regarding the total number of pediatric patients with rheumatic diagnoses evaluated at the participating institutions during the same period, a parameter necessary for calculating the percentage of pediatric patients with these disorders that require operative intervention. Other finer resolution data such as patient socioeconomic status or the type of valve (tissue or mechanical) used for valve replacement surgery also were not included in the database. Despite these limitations, the large number of patients and multiple centers contained in the PCCC database do provide more extensive characterization of cardiac surgical intervention for pediatric rheumatic diseases than has been available previously.

Conclusions

The current study investigated cardiac surgical intervention for pediatric rheumatic diseases in North America over a 21-year span. Although the study patients represent a minority of patients with pediatric rheumatic diseases, they clearly experience measurable morbidity and mortality attributable to the cardiovascular manifestations of their diseases. Additionally, pediatric rheumatic diseases account for a small but measurable proportion of the pediatric cardiac surgical volume, a proportion that is not apparently declining despite improved medical therapy for these disorders. This study highlights the need for improved understanding of the mechanisms by which rheumatic diseases damage the cardiovascular system and the need for improved recognition and management of their cardiovascular complications in children.

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References

1. Al-Abbad AJ, Cabral DA, Sanatani S, Sandor GG, Seear M, Petty RE, Malleson PN. Echocardiography and pulmonary function testing in childhood onset systemic lupus erythematosus. *Lupus* 2001;10:32–37. [PubMed: 11243507]
2. Balmer C, Fasnacht M, Rahn M, Molinari L, Bauersfeld U. Long-term follow-up of children with congenital complete atrioventricular block and the impact of pacemaker therapy. *Europace* 2002;4:345–349. [PubMed: 12408251]
3. Bitar FF, Hayek P, Obeid M, Gharzeddine W, Mikati M, Dbaibo GS. Rheumatic fever in children: a 15-year experience in a developing country. *Pediatr Cardiol* 2000;21:119–122. [PubMed: 10754079]
4. Boris JR, Hollister JR, Tyson RW, Wolfe RR. Severe aortic regurgitation with mechanical prosthetic valve replacement in pauciarticular juvenile rheumatoid arthritis. *J Rheumatol* 1998;25:1211–1214. [PubMed: 9632088]
5. Burns JC, Kushner HI, Bastian JF, Shike H, Shimizu C, Matsubara T, Turner CL. Kawasaki disease: a brief history. *Pediatrics* 2000;106:E27. [PubMed: 10920183]
6. Buyon JP, Hiebert R, Copel J, Craft J, Friedman D, Katholi M, Lee LA, Provost TT, Reichlin M, Rider L, Rupel A, Saleeb S, Weston WL, Skovron ML. Autoimmune-associated congenital heart block: demographics, mortality, morbidity and recurrence rates obtained from a national neonatal lupus registry. *J Am Coll Cardiol* 1998;31:1658–1666. [PubMed: 9626848]
7. Carapetis JR, McDonald M, Wilson NJ. Acute rheumatic fever. *Lancet* 2005;366:155–168. [PubMed: 16005340]
8. Carapetis JR, Steer AC, Mulholland EK, Weber M. The global burden of group A streptococcal diseases. *Lancet Infect Dis* 2005;5:685–694. [PubMed: 16253886]
9. Carceller A, Tapiero B, Rubin E, Miro J. Acute rheumatic fever: 27-year experience from the Montreal's pediatric tertiary care centers. *An Pediatr Barc* 2007;67:5–10. [PubMed: 17663899]
10. Cervera R, Font J, Pare C, Azqueta M, Perez-Villa F, Lopez-Soto A, Ingelmo M. Cardiac disease in systemic lupus erythematosus: prospective study of 70 patients. *Ann Rheum Dis* 1992;51:156–159. [PubMed: 1550395]
11. Chauvaud SM, Kalangos A, Berrebi AJ, Gaer AR, Acar C, Carpentier AF. Systemic lupus erythematosus valvulitis: mitral valve replacement with a homograft. *Ann Thorac Surg* 1995;60:1803–1805. [PubMed: 8787490]
12. Delgado EA, Petty RE, Malleson PN, Patterson MW, D'Orsogna L, LeBlanc J. Aortic valve insufficiency and coronary artery narrowing in a child with polyarticular juvenile rheumatoid arthritis. *J Rheumatol* 1988;15:144–147. [PubMed: 3280793]
13. Doria A, Iaccarino L, Sarzi-Puttini P, Atzeni F, Turriel M, Petri M. Cardiac involvement in systemic lupus erythematosus. *Lupus* 2005;14:683–686. [PubMed: 16218467]
14. Eronen M, Siren MK, Ekblad H, Tikanoja T, Julkunen H, Paavilainen T. Short- and long-term outcome of children with congenital complete heart block diagnosed in utero or as a newborn. *Pediatrics* 2000;106:86–91. [PubMed: 10878154]
15. Gerber MA, Baltimore RS, Eaton CB, Gewitz M, Rowley AH, Shulman ST, Taubert KA. Prevention of rheumatic fever and diagnosis and treatment of acute streptococcal pharyngitis: a scientific statement from the American Heart Association Rheumatic Fever, Endocarditis, and Kawasaki Disease Committee of the Council on Cardiovascular Disease in the Young, the Interdisciplinary Council on Functional Genomics and Translational Biology, and the Interdisciplinary Council on Quality of Care and Outcomes Research: endorsed by the American Academy of Pediatrics. *Circulation* 2009;119:1541–1551. [PubMed: 19246689]
16. Goldenberg J, Ferraz MB, Pessoa AP, Fonseca AS, Carvalho AC, Hilario MO, Atra E. Symptomatic cardiac involvement in juvenile rheumatoid arthritis. *Int J Cardiol* 1992;34:57–62. [PubMed: 1548110]
17. Gotteiner N, Mavroudis C, Backer CL, Pahl E, Duffy CE, Dalzell V, Shulman ST. Coronary artery bypass grafting for Kawasaki disease. *Pediatr Cardiol* 2002;23:62–67. [PubMed: 11922511]
18. Gunal N, Kara N, Akkok N, Cakar N, Kahramanyol O, Akalin N. Cardiac abnormalities in children with systemic lupus erythematosus. *Turk J Pediatr* 2003;45:301–305. [PubMed: 14768793]

19. Hillman ND, Tani LY, Veasy LG, Lambert LL, Di Russo GB, Doty DB, McGough EC, Hawkins JA. Current status of surgery for rheumatic carditis in children. *Ann Thorac Surg* 2004;78:1403–1408. [PubMed: 15464505]
20. Hiraki LT, Benseler SM, Tyrrell PN, Hebert D, Harvey E, Silverman ED. Clinical and laboratory characteristics and long-term outcome of pediatric systemic lupus erythematosus: a longitudinal study. *J Pediatr* 2008;152:550–556. [PubMed: 18346514]
21. Holman RC, Curns AT, Belay ED, Steiner CA, Schonberger LB. Kawasaki syndrome hospitalizations in the United States, 1997 and 2000. *Pediatrics* 2003;112:495–501. [PubMed: 12949272]
22. Izmirly PM, Rivera TL, Buyon JP. Neonatal lupus syndromes. *Rheum Dis Clin North Am* 2007;33:267–285. vi. [PubMed: 17499707]
23. Jaeggi ET, Hamilton RM, Silverman ED, Zamora SA, Hornberger LK. Outcome of children with fetal, neonatal, or childhood diagnosis of isolated congenital atrioventricular block: a single institution's experience of 30 years. *J Am Coll Cardiol* 2002;39:130–137. [PubMed: 11755298]
24. Kitamura S, Kameda Y, Seki T, Kawachi K, Endo M, Takeuchi Y, Kawasaki T, Kawashima Y. Long-term outcome of myocardial revascularization in patients with Kawasaki coronary artery disease: a multicenter cooperative study. *J Thorac Cardiovasc Surg* 1994;107:663–673. discussion 673–664. [PubMed: 8127095]
25. Kitamura S, Tsuda E, Wakisaka Y. Pediatric coronary artery bypass grafting for Kawasaki disease: 20-years' outcome. *Nippon Rinsho* 2008;66:380–386. [PubMed: 18260338]
26. Lin CH, Lee ML, Hsu RB. Cardiac surgery in patients with systemic lupus erythematosus. *Interact Cardiovasc Thorac Surg* 2005;4:618–621. [PubMed: 17670495]
27. Marijon E, Ou P, Celermajer DS, Ferreira B, Mocumbi AO, Jani D, Paquet C, Jacob S, Sidi D, Jouven X. Prevalence of rheumatic heart disease detected by echocardiographic screening. *N Engl J Med* 2007;357:470–476. [PubMed: 17671255]
28. Miyake CY, Gauvreau K, Tani LY, Sundel RP, Newburger JW. Characteristics of children discharged from hospitals in the United States in 2000 with the diagnosis of acute rheumatic fever. *Pediatrics* 2007;120:503–508. [PubMed: 17766522]
29. Moller JH, Borbas C. The Pediatric Cardiac Care Consortium: a physician-managed clinical review program. *QRB Qual Rev Bull* 1990;16:310–316. [PubMed: 2122356]
30. Moller JH, Powell CB, Joransen JA, Borbas C. The pediatric cardiac care consortium—revisited. *Jt Comm J Qual Improv* 1994;20:661–668. [PubMed: 7881516]
31. Omdal R, Lunde P, Rasmussen K, Mellgren SI, Husby G. Transesophageal and transthoracic echocardiography and Doppler examinations in systemic lupus erythematosus. *Scand J Rheumatol* 2001;30:275–281. [PubMed: 11727842]
32. Oshiro AC, Derbes SJ, Stopa AR, Gedalia A. Anti-Ro/SSA and anti-La/SS-B antibodies associated with cardiac involvement in childhood systemic lupus erythematosus. *Ann Rheum Dis* 1997;56:272–274. [PubMed: 9166002]
33. Roldan CA, Shively BK, Crawford MH. An echocardiographic study of valvular heart disease associated with systemic lupus erythematosus. *N Engl J Med* 1996;335:1424–1430. [PubMed: 8875919]
34. Tsuda E, Kitamura S. National survey of coronary artery bypass grafting for coronary stenosis caused by Kawasaki disease in Japan. *Circulation* 2004;110:II61–II66. [PubMed: 15364840]
35. Yanagawa H, Nakamura Y, Yashiro M, Ojima T, Tanihara S, Oki I, Zhang T. Results of the nationwide epidemiologic survey of Kawasaki disease in 1995 and 1996 in Japan. *Pediatrics* 1998;102:E65. [PubMed: 9832593]

Fig. 1. Pediatric Cardiac Care Consortium (PCCC) characteristics, 1985–2005. The graph depicts the annual number of PCCC centers (*line*) and cardiac operative admissions (*bars*). Only centers and admissions involving patients younger than 21 years at the time of the first procedure are depicted

Fig. 2.

Distribution of cardiac surgical procedures performed for pediatric rheumatic diseases in the Pediatric Cardiac Care Consortium, 1985–2005. The pie charts depict the percentage of the indicated type of surgical procedure for the indicated conditions. The area of each pie chart corresponds to the number of procedures for the designated condition (*n*)

Fig. 3.

Age at the time of the surgical procedure for pediatric rheumatic diseases in the Pediatric Cardiac Care Consortium, 1985–2005. The graphs depict the number of patients in each age category at the time the operative procedures were performed. Age groupings were chosen to demonstrate trends by age adequately for each of the five indicated conditions. For neonatal lupus, the data are subdivided according to initial and subsequent pacemaker insertion

Fig. 4.

Annual cardiac surgical procedures for pediatric rheumatic diseases in the Pediatric Cardiac Care Consortium (PCCC). The graph depicts the proportion of cardiac surgical procedures performed in the indicated years for pediatric rheumatic diseases including rheumatic heart disease, neonatal lupus, Kawasaki disease, systemic lupus erythematosus, and juvenile arthritis. The data were normalized to the overall annual volume of pediatric cardiac surgical procedures in the PCCC database and are restricted to patients whose first surgical admission to a PCCC site occurred before the age of 21 years

Overview of cardiac surgical procedures for pediatric rheumatic diseases in the Pediatric Cardiac Care Consortium database, 1985–2005

Table 1

Diagnosis	Patients (n)	Patients with multiple procedures (n)	Total procedures (n)	No. of procedures per patient (average, range)	In-hospital mortality (n)
RHD	160	28	200	1.3, 1–5	3
NLE	53	28	106	2.0, 1–5	2
KD	28	2	31	1.1, 1–2	1
SLE	13	1	15	1.2, 1–2	0
JRA	7	2	9	1.3, 1–2	0
Total	261	61	361	1.4, 1–5	6

Numbers reflect the patients undergoing a first surgical procedure before the age of 21 years

RHD rheumatic heart disease, KD Kawasaki disease, NLE neonatal lupus erythematosus, SLE systemic lupus erythematosus, JRA juvenile rheumatoid arthritis

Table 2

Pattern of valvular involvement in pediatric rheumatic carditis among patients in the Pediatric Cardiac Care Consortium database, 1985–2005

Valve	<i>n</i>	%
M	83	43.5
A ^a	46	24.1
M, A ^a	41	21.5
M, T	13	6.8
M, A, T	5	2.6
M, P	1	0.5
A, T	1	0.5
T	1	0.5
Total	191	100

M mitral, *A* aortic, *T* tricuspid, *P* pulmonary

^aRoss procedures accounted for 15 of the 46 aortic valve procedures and 1 of the 41 mitral/aortic valve procedures, and are not considered to have involved the pulmonary valve primarily