Aortic Dissection in Childhood and Adolescence: An Analysis of Occurrences Over a 10-Year Interval in New York State

Address for correspondence:
Charles R. Fikar
Library Manager
Caritas Health Care
Mary Immaculate Hospital
Woods Health Sciences Library
152-11 89th Avenue
Jamaica, NY 11432
crfikarmath@yahoo.com

Charles R. Fikar, MD, MSLS; Ronald Fikar, PhD
Caritas Health Care/Mary Immaculate Hospital, Woods Health Sciences Library, Jamaica, New York

ABSTRACT

Background: Rarely occurring in the pediatric and adolescent population, aortic dissection is a condition with many predisposing factors. Previous studies have suggested that congenital cardiovascular disorders are the most common predisposing conditions. Trauma-associated aortic dissection is considered a rare occurrence.
Methods: The Statewide Planning and Research Cooperative System database of New York State was used to retrieve cases of aortic dissection in persons ≤21 years old over a 10-year period. A retrospective analysis for risk or associated conditions was undertaken.
Results: Forty-five of a total of 12 142 cases of aortic dissection (0.37%) occurred in persons ≤21 years old. No patient was younger than 15 years of age. Six of the 45 died (13%). Most patients were male (37 of 45, or 82%). Contrary to previous reports, the most common associated condition was trauma (19 of 45, or 42%), with Marfan syndrome the second most common (11 of 45, or 24%). Ten of 45 adolescent and young adult patients (22%) had no apparent risk factors.
Conclusions: Traumatic aortic disruptions must be considered in children and adolescents who survive serious chest trauma. Family members of pediatric and young adult patients with trauma-associated aortic dissection may need to be evaluated for possible risk of aortic dissection.

Introduction

No prospective studies of aortic dissection in the pediatric population are available in the English-language medical literature. Limited retrospective reviews, reports, and original case studies are available on the subjects of aortic dissection and aortic aneurysm disease in children and adolescents, but very little has been written about the epidemiology and demographics of these disorders.[1–15]

Available studies indicate that from 0.67% to 3.5% of aortic dissections occur in persons ≤21 years old and that in a 1-year period in the state of New York, 2 of 5658 reported deaths in the 0- to 19-year-old population group were due to this disorder.[15]

Conditions that weaken the aortic medial layer predispose to aortic dissection.[11,15,16] Some reports have indicated that congenital cardiovascular anomalies are the most common predisposing conditions for acute aortic dissection occurring in the pediatric population.[5] Traumatic aortic dissections are usually stated to be very rare. Although causative and associated factors have been well enumerated for pediatric cases, their relative contribution to total causation in this population is speculative because of small sample size.

One major ongoing study worthy of attention in this regard is the International Registry of Aortic Dissection (IRAD), centered at the University of Michigan, Ann Arbor.[17] By using the resources of 12 referral institutions in 6 countries, a population of persons ≤21 years old is slowly being accumulated that should produce much accurate information about this disorder in this age group. From its initiation in 1996 through June 2005, the IRAD had accumulated 1351 patients, of whom 9 were ≤21 years old (0.76%).[15] At this rate of accretion, a total of about 50 pediatric patients would be expected by 2040. The IRAD does not include cases associated with trauma.

The purpose of this study was to retrospectively investigate selected aspects of the incidence, demographics, and associated or causative factors of aortic dissection in the population ≤21 years of age. Traumatic aortic dissections were not excluded from this study. Because aortic dissection rarely occurs in this age range, a large database is required to procure a meaningful sampling of affected individuals. For this reason, the Statewide Planning and Research Cooperative System (SPARCS) database of New York State was chosen as the data source. This database has been sampled for published studies involving the pediatric population[18] as well as studies pertaining to aortic aneurysms.[19]

Methods

Database Information

The SPARCS stores data for every hospital discharge, ambulatory surgical procedure, and emergency department admission in New York State that occurs in nonfederally operated hospitals and mental health institutions.[20]

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Data Collection

This was a retrospective analysis of previously collected, publicly available data obtained from SPARCS. For a 10-year period, from 1996 to 2006, age, sex, year of discharge, and all ICD9 diagnosis codes were obtained for any person ≤21 years old who also had at least 1 of the following ICD9 diagnosis codes: 441.00 (dissection of aorta, unspecified site), 441.01 (dissection of aorta, thoracic), 441.02 (dissection of aorta, abdominal), or 441.03 (dissection of aorta, thoracoabdominal).

Institutional review board approval of this study was not required because it involved only analysis of existing, publicly available data that have been recorded so they are not individually identifiable and that have been authorized by public law in the state of New York for research use.

Statistical Analysis

Standard statistical methods[21–23] for determining sample mean, median, mode, range, and standard deviation were used to summarize the basic demographics of the study group, as represented in Table 1. This basic demographic data was analyzed using SPSS 12.0 (Student Version) software (SPSS Inc., Chicago, IL). The *P* value for the percentage of male patients was determined by using the 1-sample *z* test (2-tailed), with 50% as the expected value for no sex bias. This value was calculated by the authors using the standard formula for this method.[23]

The analysis of the percentage of patients expiring in the study groups compared with the total SPARCS aortic dissection group was performed using the 2-sample *z* test (2-tailed)[23] to calculate the *P* value for the difference between population proportions. The 95% confidence interval for the difference between population proportions was calculated by standard methods.[22]

The 95% confidence intervals in Table 2 were determined through the use of the standard formula for estimating the confidence interval for population proportions.[22] These calculations were done using a Voyage 200 graphing calculator (Texas Instruments Inc., Dallas, TX).

Table 1. Demographics of Study Group[a]

Characteristic	Value (n = 45)	*P* Value[b]
Age		
Mean (years)	18.4 ± 1.93	
Median (years)	19	
Mode (years)	20	
Range (years)	15–21	
Male sex, n (%)	37 (82%)	< .001

[a] ± Values are means ± standard deviation; [b]P value calculated using the 1-sample *z* test (2-tailed).

Table 2. Risk/Associated Factors for Aortic Dissection in Persons ≤21 Years of Age Determined from Analysis of SPARCS Data

Risk/Associated Factor	Number[a] (%), n = 45	95% Confidence Interval
Male sex	37 (82%)	71%–93%
Trauma	19 (42%)	28%–57%
Marfan syndrome	11 (24%)	12%–37%
None apparent	10 (22%)	10%–34%
Aortic valve disorder	4 (8%)	0%–17%
Hypertension	3 (6.7%)	0%–14%
Mitral valve disorder	2 (4.4%)	0%–10%
Takayasu arteritis	1 (2.2%)	0%–6.5%
Fibromuscular dysplasia	1 (2.2%)	0%–6.5%

[a]Totals greater than 45 because some persons had multiple risks.

Results

The total number of cases of aortic dissection in the SPARCS database for the 10-year period was 12,142. Forty-five of these cases were ≤21 years of age (0.37%, 95% confidence interval [CI], 0.26%–0.48%]. Six of 45 died (13%), compared with 1891 deaths among the 12 142 (16%), the total number of aortic dissection patient ($P = .68$, 2-sample *z* test, 2-tailed; 95% CI, −0.12 to 0.08). Table 1 presents basic demographic information, and Table 2 presents the risk or associated factors determined from analysis of the retrieved ICD9 codes.

Discussion

Approximately 80% of persons were male. The IRAD reported that 76% of persons under age 40 with aortic dissection were male.[13] Zalzstein et al reported that 8 of a total of 13 aortic dissections in persons ≤25 years of age were male (62%).[12] No persons under age 15 were reported to have this diagnosis in our study. There was no difference in hospital mortality between the sample group and the total New York State SPARCS sample.

Trauma, generally considered a rare cause of aortic dissection in children and adolescents, was found to be the leading risk or associated condition, occurring in 19 of 45 patients (42%) in the present study. Zalzstein et al reported that in 3 of a total of 13 patients ≤25 years old (23%), aortic dissection was secondary to acute chest trauma.[12]

The pathophysiology of aortic dissection associated with trauma has not been entirely elucidated. It is thought that a combination of several types of force, including

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acceleration, horizontal deceleration, vertical deceleration, blast, compression, sudden extension of the neck, sudden traction on the shoulder, and perhaps others most likely causes the injury to the aortic wall that results in either dissection, partial tearing, or full transection.[8,24]

Traumatic aortic injury has many anatomic variants. Traumatic aortic disruption can appear as a traumatic aortic intimal tear, as a subadventitial traumatic disruption of the aorta, an entity that can be subdivided into partial, subtotal, and complete varieties, and finally as a true aortic dissection, which will be termed a trauma-associated aortic dissection.[25–28] Traumatic aortic intimal tear and subadventitial traumatic disruption of the aorta events are not the equivalent of an ordinary aortic dissection that happens to be secondary to trauma (trauma-associated aortic dissection). Trauma-associated aortic dissections occur at the rate of about 12% in persons with traumatic aortic tears or ruptures.[29,30] The ICD9 diagnostic coding system does not accommodate these anatomic and pathologic distinctions. For the purposes of this study, it will be assumed that all ICD9-coded aortic dissections occurring with severe trauma represent true aortic dissections (trauma-associated aortic dissections).

Motor vehicle, pedestrian, and bicycle accidents, as well as sports injuries and falls, would seem to be the more common major inciting traumatic events. Persons whose aortas are already predisposed to dissection because of weakening of medial elastic fibers would presumably be at higher risk for trauma-associated aortic dissection.[24]

It would seem prudent to examine patients presenting with trauma-associated aortic dissection for disorders predisposing to nontraumatic aortic dissection. If such a condition is found and is heritable, family members would then need to be investigated.[15] Whether it would be necessary and cost effective to evaluate family members of all patients with trauma-associated aortic dissection is unknown.

Ten of 45 persons in this study (22%) had no apparent causal or associated disorder. If the 19 trauma patients are omitted, 10 of 26 persons (38%) had no apparent risk factors. It would have been prudent for the first-degree relatives of these persons to have been evaluated for aortic root enlargement because many conditions that predispose to aortic dissection are inherited and have no discernable phenotypic features except for an intrinsically weakened aortic media.[15]

Certain disorders of connective tissue are well known to predispose to aortic dissection. Eleven of the 45 persons in this study (24%) had Marfan syndrome. The IRAD reported that 50% of their patients under age 40 had this syndrome.[13] If the 19 trauma patients in this study are omitted, 11 of 26 persons (42%) had Marfan syndrome, similar to that in the IRAD study, which, as previously stated, did not include trauma-associated cases. Zalzstein et al reported that 4 of a total of 13 cases (31%) in persons ≤25 years old had Marfan syndrome.[12] Other known connective tissue disorders associated with this condition in persons ≤21 years old were not apparent in this study.[15,31]

Congenital cardiovascular anomalies were associated with 7 of 45 patients (16%) in this study, including fibromuscular dysplasia and aortic and mitral valve disorders. If the 19 trauma patients in this study are omitted, 7 of 26 persons (27%) in our study had congenital cardiovascular anomalies. Both of the cases of mitral valve disorders were in persons diagnosed with Marfan syndrome, as were 2 of the 4 cases of aortic valve disorders. Zalzstein et al reported that 5 of 13 persons ≤25 years old (38%) had some form of congenital cardiovascular disorder.[12]

Hypertension was diagnosed in 3 of the 45 persons in this study (6.7%). None of the cases of Zalzstein were reported to be hypertensive.[12] The IRAD reported that 34% of their patients under the age of 40 were hypertensive.[13] Again, if the 19 trauma patients in this study are omitted, 3 of 26 persons (12%) were hypertensive.

There were many limitations to this study. The ICD9 coding system cannot replace a medical chart review. It has been stated that the accuracy of coding, at least for surgical diagnoses, is approximately 90%.[32] Because there are no patient identifiers, readmissions and multiple admissions were not excluded from the data.

Despite many limitations, it would seem that because of the large size of the database and that the reporting of such data is mandatory, the hospitalization data for a 10-year period in the state of New York is providing useful and significant information on a topic that otherwise is extremely difficult to study.

Conclusion

In summary, it is clear that chest trauma is a leading cause of aortic dissection or disruption in young persons. Marfan syndrome and congenital cardiovascular anomalies are also common predisposing conditions. If a trauma-associated aortic dissection patient has a known predisposing condition, it is suggested that first-degree kin undergo screening. It is unknown if the families of patients with trauma-associated aortic dissection with no apparent predisposing factor need screening. This question could be a topic for additional research.

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