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Original Article

Aortic dissection in children and young adults: diagnosis, patients at risk, and outcomes

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Abstract *Objective:* To heighten the awareness of pediatricians and pediatric cardiologists to aortic dissection, a potentially dangerous medical condition. *Methods:* We reviewed the charts of 13 patients, seen in four medical centers, who suffered acute or chronic aortic dissection over the period 1970 through 2000 whilst under the age of 25 years. *Results:* There were seven male and six female patients, with the mean age at diagnosis being 12.1 years, with a range from one day to 25 years. Congenital cardiac defects were present in five patients, and Marfan syndrome in four. In three of the patients with congenital cardiac defects, aortic dissection developed as a complication of medical procedures. In three patients, dissection followed blunt trauma to the chest. We could not identify any risk factors in one patient. The presenting symptoms included chest pain in four patients, abdominal pain and signs of ischemic bowel in two, non-palpable femoral pulses in one, and obstruction of the superior caval vein in one. Angiography and magnetic resonance imaging were the main diagnostic tools. Overall mortality was 38%. Only six patients had successful surgical outcomes. *Conclusion:* Due to the rarity of aortic dissection a high index of suspicion is required to reach the diagnosis in a timely manner. It should be considered in young patients complaining of chest pain in association with Marfan syndrome, anomalies of the aortic valve and arch, and chest trauma.

Keywords: Aortic aneurysm; Marfan syndrome; chest trauma

Aortic dissection is rare in children and young adults. In adults over 50, progressive arteriosclerosis and chronic hypertension are the most common risk factors. The risk factors most commonly associated with this condition in the young are inherited disorders of the connective tissues, congenital cardiac disease, severe trauma, use of cocaine, and chronic hypertension.[1–6] We describe here 13 patients who were diagnosed and treated for aortic dissection in four medical centers over a period of 30 years. We have reviewed the risk factors, mode of diagnosis, treatment, and outcome, emphasizing the need to increase awareness of the condition so as to facilitate early diagnosis and ensure optimal management.

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Methods

We saw 13 children and young adults with aortic dissection between 1970 and 2000 in four medical centers, namely Soroka University Medical Center, Tel Aviv Medical Center, Children's Hospital, Toronto, and Toronto General Hospital. We identified the patients by diagnostic code from the archives of the hospital and the departments of cardiology, extracting their relevant clinical details (Table 1).

Results

The mean age of the 13 patients at diagnosis was 12.1 years, with a range from 1 day to 25 years. There

Table 1. Demographic and clinical data of patients diagnosed and treated for aortic dissection.

Patient	Age at onset	Gender	Symptoms	Diagnosis	Site of dissection	Imaging	Treatment	Outcome
1	1 day	M	Bowel ischemia	TOF, PA, PAD stent placement	Descending aorta	Angiography, echo	Conservative	Expired
2	2 y	M	Bowel ischemia	Cannulation for TOF	Ascending aorta	Echo	Conservative	Expired
3	8 m	F	Diminished pedal pulses	PAD, occluding device	Descending aorta	Angiography	Conservative	Resolved
4	15 y	M	None	Marfan	Ascending aorta	MRI	Surgery	Resolved
5	17 y	M	Chest pain	Marfan	Ascending aorta	Angiography	Surgery	Resolved
6	12 y	M	Chest pain	Blunt trauma	Ascending aorta	TEE	Surgery	Resolved
7	7 y	F	None (coma)	Road accident	Ascending aorta	TEE	Conservative	Expired
8	14 y	M	Absent femoral pulses	Road accident	Descending aorta	Angiography	Surgery	Resolved
9	19 y	M	SVC syndrome	Aortic valve stenosis after valve replacement	Ascending aorta	Angiography	Surgery	Resolved
10	25 y	F	Chest pain	Marfan	Ascending/descending	TEE	Surgery	Resolved
11	21 y	M	Chest pain	Marfan	Ascending/transverse	TEE	Surgery	Resolved
12	8 y	F	Chest pain	Coarctation	Ascending aorta	None		Expired
13	17 y	F	Chest pain		Ascending aorta	None		Expired

Abbreviations: TOF: Tetralogy of Fallot; PA: pulmonary atresia; CBP: cardiopulmonary bypass; PAD: patency of arterial duct; MRI: magnetic resonance imaging; TEE: transesophageal echocardiography

were seven males and six females. The dissection was acute in twelve of the patients. In six patients, there was acute onset of chest pain, while two had abdominal pain or signs of bowel ischemia, and one patient had neck swelling and signs of the superior caval venous syndrome. Femoral pulses were absent in two patients. One patient with chronic dissection due to Marfan syndrome was asymptomatic. The diameter of the aortic root prior to dissection was in the upper range in all patients with Marfan syndrome, specifically between 56 and 61 mm, and none had a family history of dissection. Findings consistent with dissection were seen on echocardiography. A congenital cardiac defect was present in five patients, with tetralogy of Fallot present in two, patency of the arterial duct in one, aortic valvar stenosis in one, and aortic coarctation in the other. Marfan syndrome had been diagnosed in four patients. In three patients, the dissection followed blunt trauma to the chest during road accidents, while the final patient had no previous known risk factor.

In three patients with congenital cardiac defects, aortic dissection was a complication of a medical procedure. The first of these patients had undergone implantation of a stent in the arterial duct on the first day of life in the setting of Tetralogy of Fallot with pulmonary atresia. The stent embolized to the descending aorta, and aortic dissection occurred during attempts at retrieval. The second patient developed dissection of the ascending aorta when cannulation of the superior caval vein was performed during a cardiopulmonary bypass procedure. The third patient developed dissection of the descending aorta following migration of an occluding device placed to block a large patent arterial duct. The device embolised into the descending aorta when attempts were made to retrieve it.

The diagnosis of dissection had been made in 10 patients, in five by aortic angiography, in four by transoesophageal echocardiography, and in one by magnetic resonance imaging. The overall rate of mortality was 38%. Surgical repair was performed in seven patients, all of whom survived. Three of the four patients died in whom no surgical repair was attempted. In the first and second of these patients, the surgical risk was considered too great. In the seventh patient, a conservative therapeutic approach was adopted because the neurological state of the patient left no hope for recovery. In the eleventh and twelfth patients, the diagnosis of acute dissection was missed, being made only at autopsy.

Sample cases

Patient #3

Our third patient, aged 8 months, was admitted for elective cardiac catheterization. She had a large patent arterial duct, had failed to thrive, and was treated with digoxin. Cardiac catheterization was performed under general anaesthesia. Angiography revealed the diameter of the duct to be 4 mm, so occlusion was attempted using a 7 mm Gianturco/Grifka pouch. A small residual leak was seen immediately afterwards, but was expected to close spontaneously. The patient

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remained in the hospital overnight due to vomiting. The next morning, her femoral pulses were diminished, with a loud continuous murmur raising suspicion of embolization, a condition that was confirmed by X-ray. The patient was returned to the catheterization laboratory for retrieval of the device. After considerable manipulation, the device was retrieved transvenously using a basket catheter, but it would no longer pass through the duct. A wire was introduced through an arterial route for aortography, but could not be advanced, raising suspicion of aortic dissection. This was confirmed by angiography, which showed the lesion to extend from the celiac axis to the level of the tenth thoracic vertebra. A repeat injection 90 min later did not show further extension of the dissection. The patient was taken to the operating room for removal of the device and ligation of the arterial duct. The dissection of the aorta was managed conservatively. Follow-up magnetic resonance imaging showed thrombosis of the dissection, with no aneurysmal dilation of the aorta.

Patient #10

Our tenth patient was a 25-year-old female with Marfan syndrome. She developed severe central chest pain while talking on the phone with a friend. The pain continued for about 5 min, after which it became slightly less intense. She went to bed, but when the discomfort persisted, she went to the hospital emergency room where she was seen by a family physician who suggested that she discontinue the erythromycin that she was taking for a common cold. She went home, but returned to the emergency room 13 hours later because the pain increased in severity. A chest X-ray, and a chest computerized tomographic scan, revealed no abnormalities. Intense pain then developed in the interscapular area. The patient was treated with morphine and was sent to Toronto General Hospital for further evaluation. She underwent an emergency transoesophageal echocardiography that demonstrated an aortic dissection in the ascending aorta extending around the arch to the descending aorta. There was evidence of communication between the true and false lumen above the aortic valve, and moderate to severe aortic insufficiency. The patient was taken to the operating room where the dissection was successfully repaired.

Patient #12

The twelfth patient had been followed with a bifoliate aortic valve and repair of discrete aortic coarctation. The surgical procedure had been uneventful, and yearly follow-up visits at the cardiology clinic showed no stenosis or regurgitation of the aortic valve. At the age of 8 years, she complained of increasing fatigue. Echocardiography showed moderate stenosis at the site of coarctation, that had been repaired by a subclavian flap. The ascending aorta was dilated, without change over a period of 5 years, with a maximal diameter of 42 mm. She developed acute chest pain, and was referred to an emergency room. The pain gradually disappeared, and she was discharged with a diagnosis of musculoskeletal chest pain and treated with analgesics. The next morning she was found dead in her bed. An autopsy revealed dissection of the ascending aorta.

Discussion

Aortic dissection is rare in children and young adults, occurring most commonly in the fifth to seventh decade of life. The known incidence is 5–10 cases per million of the population.[1] Hirt et al.[7] reviewed 505 cases of dissecting aneurysm, of which only 7 were under the age of 20 years, and none was younger than 14 years.

The predisposing conditions for aortic dissection can be divided into 4 categories:

- Degenerative and inflammatory vascular conditions.
- Congenital cardiovascular anomalies, including chronic systemic hypertension.
- Trauma to the aortic wall.
- Miscellaneous activities, including use of cocaine, weight lifting, pregnancy, and so on.

Marfan syndrome is the most common inherited connective disorder in which an abnormality of fibrillin leads to progressive weakness of the aortic wall.[3] Other implicated disorders of connective tissue include the Ehlers-Danlos syndrome,[8] Osteogenesis imperfecta,[9] and Turner's and Noonan's syndromes. Patients with inflammatory processes such as Takayasu's arteritis, giant cell arteritis, and so on, have also been reported with aortic dissection.[2]

There appears to be a close association between bifoliate aortic valve and aortic dilation, resulting in formation of aneurysms and dissection. The aorta of patients with aortic coarctation is considered inherently predisposed to dissection, regardless of whether it is treated by balloon angioplasty or surgical repair.[10,11]

Chest trauma to the aortic wall has been recognized as a risk factor for aortic dissection for years, but recently weight lifting,[12] and use of cocaine or crack by young persons,[13] has been added to the list. It has been speculated that the very high blood pressure generated during the lifting and the pharmacologic effect of cocaine may cause tearing of the aortic intima. Aortic dissection also has been reported in

previously healthy children and adolescents with no known risk factors.[14] Any mechanism that weakens the aortic wall, especially the aortic lamina media, can lead to higher wall stress and the induction of aortic dilation and aneurysmal formation, eventually resulting in aortic dissection or rupture. Medial abnormalities in the ascending aorta, and in the aorta adjacent to sites of coarctation, have been found in biopsies from a variety of congenital cardiac diseases.[15] Dissection occurs when blood dissects between the middle and outer two thirds of the media, creating a blood-filled canal within the aortic wall.[16] The process usually occurs in the ascending aorta within 10 cm of the aortic valve, where an intimal tear is usually observed. This dissecting intramural hematoma may rupture into the pericardial, pleural, or peritoneal cavities. Forward extension of the hematoma may cause partial or total obstruction of the carotid, renal, mesenteric, iliac, or spinal arteries, with subsequent catastrophic complications. Backward extension may cause obstruction of the coronary arteries, or malfunction of the aortic valve.[17]

The fact that we identified only 13 patients with aortic dissection over a period of 30 years in four tertiary medical centers highlights the rarity of this condition in children and young adults. Over nine-tenths of the patients had a major risk factor for aortic dissection. Marfan syndrome was diagnosed in four patients, out of 218 patients followed during the period of study, congenital cardiac malformations in five, out of 3152 patients with a bicuspid aortic valve, and 2272 patients with aortic coarctation, and chest trauma in three. In only one patient were we unable to find any known risk factor. In three of the five patients with congenital cardiac malformations, aortic dissection occurred as a complication of a medical procedure. This reflects the increasing weight of interventional procedures in routine pediatric cardiologic care.

Surgical repair was performed in seven patients, all of whom survived. Of the four patients in whom we adopted a conservative approach, however, three died. This approach was chosen because the risk of a surgical procedure was too great. Clearly, whenever feasible, a surgical approach is superior to medical therapy, and increases the chances for survival. Unfortunately, two patients died suddenly before the diagnosis of aortic dissection could be reached. The twelfth patient was known to have aortic coarctation, while the other had no predisposing condition. Although a surgical approach is recommended, no patient with aortic dissection should be considered cured following successful surgery, and follow up is mandatory.

The main challenge in managing acute aortic dissection is to suspect and diagnose the disease as early as possible. Because of its rarity in the younger population, prompt diagnosis is often delayed. The outcome can then be catastrophic. The cases presented here illustrate the importance of a high index of suspicion in any young patient with chest pain, back pain and/or decreased perfusion of the lower limbs in the presence of known predisposing factors of aortic dissection. One also needs to be aware of less common presentations. Chest pain is often atypical or may be absent. Up to one-fifth of patients with acute aortic dissection may present with syncope without a history of typical pain.[18]

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