Successful endovascular treatment of type B aortic dissection in children. First-in-man experience

Ivo Petrov, MD, PhD, FESC, FACC
Head Cardiology, angiology electrophysiology
Acibadem City Clinic, Sofia, Bulgaria

petrovivo@hotmail.com
Disclosure Statement of Financial Interest

I, Ivo Petrov

DO NOT have a financial interest/arrangement or affiliation with one or more organizations that could be perceived as a real or apparent conflict of interest in the context of the subject of this presentation.
Introduction

• Acute aortic dissection (AD) is extremely rare among the population under 18 years of age.

• An average of 0.67% (range, 0.37% to 3.5%) of all aortic dissections occur in young patients, predominantly in males.

• The data regarding patient characteristics and treatment management of such aortic pathology are sparse and limited to small populations and few case reports.

Ao dissection in childhood.
Literature revision:

• Risk factors among young patients:

• connective tissue disorders (Marfan and type IV Ehlers-Danlos), congenital cardiovascular disease - aortic coarctation, aortic valvular stenosis, unicuspid/ bicuspid aortic valve, patent ductus arteriosus, trauma, drug abuse, weight lifting and hypertension.

5. Keiji Uchida, Acute aortic dissection occurring during the butterfly stroke in a 12-year-old boy
6. C J Hogan An aortic dissection in a young weightlifter with non-Marfan fibrillinopathy
Table 5. Genetic Syndromes Associated With Thoracic Aortic Aneurysm

<table>
<thead>
<tr>
<th>Genetic Syndrome</th>
<th>Common Clinical Features</th>
<th>Genetic Defect</th>
</tr>
</thead>
<tbody>
<tr>
<td>Marfan syndrome</td>
<td>Skeletal features (see text)</td>
<td>FBN1 mutations*</td>
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<tr>
<td></td>
<td>Ectopia lentis</td>
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<td></td>
<td>Dural ectasia</td>
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<tr>
<td>Loes-Dietz syndrome</td>
<td>Bifid uvula or cleft palate</td>
<td>TGFBR2 or TGFBR1 mutations</td>
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<tr>
<td></td>
<td>Arterial tortuosity Hypertelorism</td>
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<tr>
<td></td>
<td>Skeletal features similar to MFS Craniostenosis</td>
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<td></td>
<td>Aneurysms and dissections of other arteries</td>
<td></td>
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<tr>
<td>Ehlers-Danlos syndrome, vascular form</td>
<td>Thin, translucent skin</td>
<td>COL3A1 mutations</td>
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<td></td>
<td>Gastrointestinal rupture</td>
<td></td>
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<td></td>
<td>Rupture of the gravid uterus</td>
<td></td>
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<tr>
<td></td>
<td>Rupture of medium-sized to large arteries</td>
<td></td>
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<tr>
<td>Turner syndrome</td>
<td>Short stature</td>
<td>45, X karyotype</td>
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<td></td>
<td>Primary amenorrhea</td>
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<td></td>
<td>Bicuspid aortic valve</td>
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<tr>
<td></td>
<td>Aortic coarctation</td>
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<tr>
<td></td>
<td>Webbed neck, low-set ears, low hairline, broad chest</td>
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</tr>
</tbody>
</table>

* The defective gene at a second locus for MFS is TGFBR2 but the clinical phenotype as MFS is debated.

Table 4. Gene Defects Associated With Familial Thoracic Aortic Aneurysm and Dissection

<table>
<thead>
<tr>
<th>Defective Gene Leading to Familial Thoracic Aortic Aneurysms and Dissection</th>
<th>Contribution to Familial Thoracic Aortic Aneurysms and Dissection</th>
<th>Associated Clinical Features</th>
<th>Comments on Aortic Disease</th>
</tr>
</thead>
<tbody>
<tr>
<td>TGFB2 mutations</td>
<td>4%</td>
<td>Thin, translucent skin</td>
<td>Multiple aortic dissections documented at aortic diameters &lt;5.0 cm</td>
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<tr>
<td></td>
<td></td>
<td>Arterial or aortic tortuosity</td>
<td></td>
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<tr>
<td></td>
<td></td>
<td>Aneurysm of arteries</td>
<td></td>
</tr>
<tr>
<td>MYH11 mutations</td>
<td>1%</td>
<td>Patent ductus arteriosus</td>
<td>Patient with documented dissection at 4.5 cm</td>
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<tr>
<td>ACTA2 mutations</td>
<td>14%</td>
<td>Livedo reticularis</td>
<td>Two of 13 patients with documented dissections &lt;5.0 cm</td>
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<tr>
<td></td>
<td></td>
<td>Iris fioccoli</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Patent ductus arteriosus</td>
<td></td>
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<tr>
<td></td>
<td></td>
<td>Bicuspid aortic valve</td>
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</tr>
</tbody>
</table>

ACTA2 indicates actin, alpha 2, smooth muscle aorta; MYH11, smooth muscle specific beta-myosin heavy chain; and TGFBR2, transforming growth factor-beta receptor type II.
• Several cases of Ao dissection with fatal end
• Several cases treated surgically (most unfavourable outcome)
• 1 case of iatrogenic dissection treated with stent graft
• No one spontaneous type B dissection treated by endovascular means

2. Hatzaras IS, Role of exertion or emotion as inciting events for acute aortic dissection. Am J Cardiol. 2007;100:1470–1472.
3. Keiji Uchida, Acute aortic dissection occurring during the butterfly stroke in a 12-year-old boy
Case reports

• In the period between 2014 and 2017 we had two patients under the age of 18 with complicated aortic dissection which we treated by endovascular means.

Total: 273
• F.E.V. 15-year-old child presented at the Emergency Department (ED) of a municipal hospital with sudden onset of back and chest pain, peak of systolic pressure 200/130 mmHg and numbness in the left leg.

• Past medical history: surgical correction of PDA at the age of 6 months. No family history of connective tissue disorders.

• CT scan: was AD Stanford B with severe compression of the true lumen and origin of the intimal tear just distal to the left subclavian artery. The dissection reached the aortic bifurcation and pervaded the left common iliac artery (total occlusion with unsatisfactory collateral flow). Superior mesenteric and left renal arteries were totally occluded causing ileus and elevated renin levels with extremely resistant to treatment hypertension).
MSCTA at admission:

- TBAD
- Full obliteration of vessel lumen at left renal, Superior mesenteric, left iliac arteries
• At hospitalization he had symptoms of critical end-organ ischemia (malperfusion syndrome is the second leading cause of death in TBAD):
  • ileus (due to the occlusion of the superior mesenteric artery and subocclusive celiac trunk),
  • severe limb ischemia, paresis and necrosis of the left lower limb (with peak creatine phosphokinase [CPK] value of 20,000 IU/L during the course of the clinical episode),
  • occlusion of the left renal artery leading to uncontrolled arterial hypertension reaching values of 220/140 mmHg.

• A multidisciplinary heart and vascular team discussion:
  • Due to the underlying life-threatening emergency condition, conservative medical treatment was not considered as a reasonable treatment option. Radical surgical treatment was considered too risky. A multi-stage endovascular treatment strategy was accepted as the most reasonable.

1. Petrov et al., Successful EVT of type B Aortic dissection in a 15-year-old child, Cor Et Vasa (2016)
Endovascular treatment (1-4)

- Restoring the flow in the true lumen and in the occluded branches and to overcome the life threatening ischemia was crucial for the survival of the boy. Despite lack of evidence or previous experience, decision for endovascular multi-staged treatment was taken as the least risky procedure.

With limited surgical femoral approach abdominal aortic severe compression was treated with a Zenith dissection 36/186 mm implantation resulting in full flow restoration.

Right radial approach 6 Fr:
- Left iliac artery fenestration Trail Blazer (Medtronic) microcatheter over a .035”Glidewire-Terumo), ballooning (Admiral 8.0/80mm, 8 atm), flow restauration, no residual gradient. Co-Chr stent (Valeo 5.0/20mm) in the SMA.
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Flow centralized in the true lumen of abdominal aorta, visceral vessels and both iliac arteries. No false lumen visible on these levels.
Endovascular treatment (3-4)

• During the postprocedural period, acute renal failure with tendency for oliguria, elevated creatinine, and resistant hypertension and elevated rennin levels were noted.

• Because of these findings, on the 20th postprocedural day, recanalization and stenting of the left renal artery were carried out.

• Left radial acces 6 Fr, JR 6Fr GC. Whisper wire across the struts of Zenith Dissection, crossing the dissection membrane, Dynamic Renal 5.5/18mm implantation

1. Petrov et al., Successful EVT of type B Aortic dissection in a 15-year-old child, Cor Et Vasa (2016)
This third stage of the endovascular treatment was performed 33 days after the first procedure.

The procedure was carried out under general anesthesia with a limited left femoral surgical approach and bilateral radial approaches.

Diagnostic “Pigtail” catheters were positioned, the first in the aortic arch for diagnostic angiography purposes and the second in the left subclavian ostium to mark the target zone to be covered.

After lowering the blood pressure to 80/50 mmHg, implantation of a Zenith Alpha stent-graft (Cook Medical) over a stiff Lunderquist wire (Cook Medical) was performed.

Balloon molding (with a compliant balloon Reliant (Medtronic Vascular) to ensure that complete proximal apposition and isolation of the entry of the dissection.

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Uneventful Follow-up

- The one, 6, 12 and 24 months follow-up were remarkable with lack of clinical events and full physical and psychological restoration. CT revealed homogenously contrasted thoracic and abdominal aorta, full obliteration of the false lumen with preserved blood flow in the previously affected arteries (left iliac, left renal artery, SMA). Normal blood samples results incl. creatinin and renin levels. The genetics samples revealed ACTA 2 mutation associated with elastopathies.
Case report

Successful endovascular treatment of type B aortic dissection in a 15-year-old child

Ivo Petrov a,*, Anna Kaneva-Nencheva b, Elisaveta Levunlieva b, Kamelia Genova b, Iskren Garvanski a, Georgi Konstantinov c, Gloria Adam d

a City Clinic Sofia University Hospital, Cardiology Department, Bulgaria
b National Heart Hospital, Pediatric Cardiology Department, Bulgaria
c National Heart Hospital, Vascular Surgery, Bulgaria
d City Clinic Sofia University Hospital, Diagnostic Imaging Department, Bulgaria

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ABSTRACT

Aortic dissection is a rare condition in the pediatric and young adult population [1]. Here, we present an unusual case of a 15-year-old male patient with sudden onset of chest and back pain and numbness in the left leg. Chest and abdominal CT revealed Stanford type B (DeBakey type III) aortic dissection, leading to subocclusive stenosis of the superior
• False lumen completely resolved
• No excessive growth of the aorta
False lumen completely resolved. No excessive growth of the aorta.
Case report-2

- The second patient was 17-year-old child presenting in the ED with severe uncontrollable chest and back pain and numbness of both legs, extremely resistant to medical treatment hypertension (reaching 220/140mmHg, on i.v. medication and signs of renal failure (Creatinine level reached 240mmol/l)).

- No history of previous cardiovascular disorders in childhood
- The CT-scan showed an aortic dissection Stanford type B, with entry 11cm distally from the left subclavian artery with false lumen practically occluding the aorta on the level of the diaphragm.
Endovascular treatment TBAD:

- Vascular access: left radial approach 5 Fr
- Right femoral limited surgical approach
- During the procedure full obliteration of the true lumen was established. Retrograde recanalization from femoral approach was not successful.
- Antegrade recanalization of the true lumen was achieved by the radial approach using triple telescoping supportive technique with 6 Fr long hydrophilic sheath (Checkflo), Multipurpose 5fr catheter and straight .035" Glidewire (Terumo). Externalization of the wire through the femoral site (“Through and through” technique) and gentle balloon predilation (with 6.0/60mm Admiral balloon), 18 Fr femoral sheath was introduced.
- Stentgraft Zenith 28/202mm was implanted covering the entry tear and telescoping with two additional Zenith-Dissection 36/120mm stents for additional centralization of the flow without any balloon postdilation (“petticoat” maneuver).
Completion angiogram:
Centralization of flow in the aorta, restored branches’ flow
- The patient survived and was discharged 23 days later in excellent clinical condition.
- The renin and blood pressure levels normalized as early as 14 days after the endovascular procedure during the hospital stay.

- The patient had no major complaints up to 6 months after the endovascular treatment and the CT scans showed optimal isolation of the false lumen and full restoration of flow in the true lumen.
CTA F-up September 2017
Conclusion:

• Several cases of Ao dissection in childhood with fatal end described in the literature
• Several cases treated surgically (most with unfavorable outcome)
• Our cases are the first described successful totally endovascular repair of a spontaneous aortic dissection of children in the world
• Early and midterm follow-up is favorable
• Strict follow-up will show if further interventions will be necessary in the future and what will be the behavior of endovascular devices in pediatric population (more liberal oversizing could probably improve the late outcome)
• The potential of endovascular therapy of the aorta in childhood has to be checked in larger series

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