Aortic dissection in a nine-year-old boy with Loes-Dietz syndrome

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A nine-year-old boy was admitted to the paediatric emergency room for inter-scapular chest discomfort since the previous day before accidental chest trauma. On physical examination, the patient presented with oesophageal hypertrophy, cranio-osteopathy, scapho-ovalia, bifid uvula, pec-trocranium, joint laxity and hypermobility, club foot, velvety and translucent skin, possibly compatible with a Loes-Dietz syndrome. The chest X-ray showed an enlarged mediastinal shadow. Bi-dimensional echocardiography revealed severe aortic root and sino-tubular junction dilatation respectively 5.8cm (age:4.8-8.9) and 5cm (age:4.8-2.6). The aortic valve was tricuspid with moderate regurgitation and the aortic annulus was 2.7cm (age:2.3-3). A suspicion of internal flap at the aortic isthmus with a pseudo-aneurysm of the wall was followed by a CT scan confirmation of an aortic dissection. Coarctation 7.2cm x 4.2cm.

The patient was immediately transferred to the OR, where he underwent an emergency replacement of the aortic isthmus with a 16mm Dacron graft on a mild hypothermic (32°C) cardiopulmonary bypass with left lateral thoracotomy. The patient was extubated on the first postoperative day and discharged from ICU on the 4th postoperative day. His recovery after surgery was uneventful, and he discharged home on postoperative day 12, on beta blocker and losartan therapy. During the hospital stay a genetic recognition confirmed the diagnosis of Loes-Dietz type 1 with mutation of the TGFBR1 gene. The patient was asked to avoid vigorous physical activities and he is now scheduled for a elective replacement of the ascending aorta.

False aneurysm origination from the proximal anastomosis of a right ventricular to pulmonary artery shunt following staged repair of Hypoplastic Left Heart Syndrome

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The stented plication has become the treatment of choice for newborns presenting with hypoplastic left heart syndrome at many centres. A modification of the classic Norwood I procedure involves placing a shunt from the right pulmonary artery to the right atrium in one stage of cardiac surgery. At the second stage, the right atriotomy is enlarged via a right atrial appendage arteriotomy and a shifted extracardiac conduit is used. This is followed by a Norwood II procedure at the third stage. In this presentation we report on a 2.5 year old boy who presented with a hypoplastic left heart syndrome and was operated on at the age of 5 months. He presented with a right to left shunt with a flow of 650 ml/min. Two Norwood procedures and a Fontan operation were performed.

Serratoa mesoracis is a rare cause of mediastinalts and bloodstream infection in children. The recurrent infections, caused by Serratoa mesoracis in the presenting patient may have been triggered by the polytetrafluorethylene shunt, leading to the deficient anastomosis. The presented technique of clipping the shunt and leaving parts of the materials in situ is a standard approach in our clinic. We asked ourselves whether to change our strategy towards a complete resection of any shunt material. On the one hand, clipping the shunt is simple, and fast. There is a low risk of bleeding, and no additional injury to the systemic right ventricle. On the other hand, leaving foreign material may increase the risk of infections.

At our institution, 37 patients who had undergone a Norwood operation with a right ventricular to pulmonary artery shunt had completion of the Fontan circula-

tion at the present time. The presented case was the single patient, suffering from this complication in our experience. Therefore, we do not see a need of changing our current strategy.