Learning from experience (Congenital): Room Jura **(14:00-15:00)**

Aortic dissection in a nine-year-old boy with Loeys-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 after accidental chest trauma. On physical examination, the patient presented with orbital hypertelorism, craniostenosis, scaphocephaly, bifid uvula, pectus excavatum, joint laxity and hypermobility, club foot, velvety and translucent skin, possibly compatible with a Loyes-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 (Z score +8.39) and 5cm (Z score +8.26). The aortic valve was tricuspid with moderate regurgitation and the aortic annulus was 2.7cm (Z score +2.94). A suspicion of intimal flap at the aortic isthmus with a pseudo-aneurism of the wall was followed by a CT scan confirmation of an isthmic aortic dissection with pseudoaneurism (7.2cm x 4.2cm).

The patient was immediately transferred to the our OR where he underwent an emergent replacement of the aortic isthmus with a 16mm Dacron graft on a mild hypothermic (30C°) cardio-pulmonary bypass through 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 was discharge home on postoperative day 12, on beta blocker and losartan therapy. During the hospital stay a genetic recognition confirmed the diagnosis of Loyes-Dietz type 1 with mutation of the TGFBR1 gene. The patient was ordered to avoid vigorous physical activities and he is now scheduled for an elective replacement of the ascending aorta.

Learning from experience (Congenital): Room Jura **(14:00-15:00)**

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 staged palliation 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 ventricle to the pulmonary arteries. At the second stage, the shunt is taken down, and a Glenn anastomosis is performed. Completion by a total cavopulmonary connection is performed as the third and last step of definitive palliation.

A three-year-old boy who had had undergone the Norwood I operation, and the Glenn operation, presented with a cervical pulsating tumor prior to the operation for total cavopulmonary connection. At the Glenn operation, the right ventricle to the pulmonary artery shunt was closed with a clip proximally, and the distal part was resected. Follwing the Glenn operation, the child had had recurrent deep sternal infections caused by Serratia marcescens. Cardiac catheterization showed a huge false aneurysm originating from the proximal shunt anastomosis (Figure 1a).

Computed tomography confirmed the diagnosis (Figure 1b). The bleeding after resternotomy was managed by initiating cardioplumonary bypass via the groin vessels. Cerebral air embolies were prevented by systemic application potassium, to achieve cardioplegic arrest during chest opening. A defect of 5mm length at the proximal suture line of the shunt was closed tentatively using enforced 5/0 prolene sutures (figure 2a). Looking towards the head, the cervical extend of the aneurysm, which caused the pulsating cervical tumour, was visible (figure 2b). During rewarming, the shunt was completely removed and the defect at the right ventricle closed using enforced 4/0 sutures. After shunt was confirmed free from infection, the total cavopulmonary connection was performed after three days.

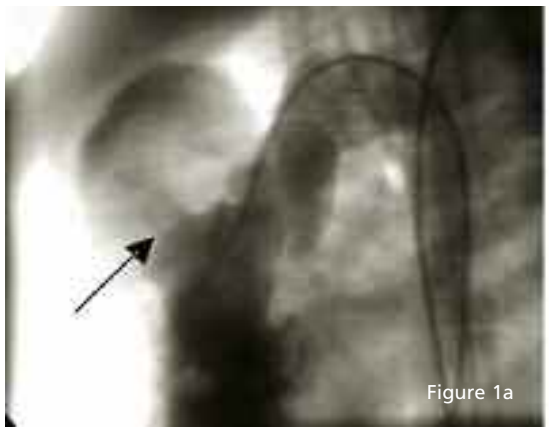
Figure 1a

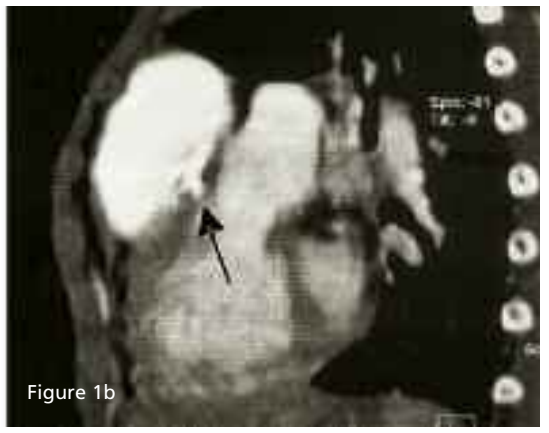
Figure 1b

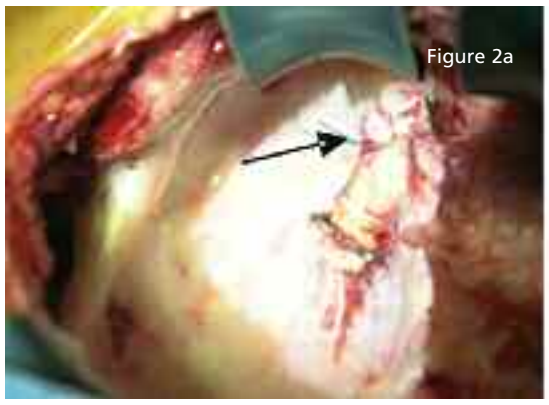
Figure 2a


Figure 2b

Serratia marcescens is a rare cause of mediastinitis and bloodstream infection in children. The recurrent infections, caused by Serratia marcescens in the presented patient may have been triggered by the polytetrafluorethylene shunt, leading to the dehiscent anastomosis. The presented technique of clipping the shunt and leaving parts of the material in situ is the standard approach at our clinic. We asked ourselves whether to change our current 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 had undergone a Norwood operation with a right ventricular to pulmonary artery shunt, had completion of the Fontan circulation 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.

AC syndrome during infant cardiac surgery

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During technically uncomplicated surgery venous drainage became problematic, therefore CPB flow (60 to 120ml kg-1 min-1) and body temperature (22.6° to 31.5°C) were reduced and large fluid volumes were administered. A marked positive balance at the end of CPB resulted 167.4 to 270 ml kg-1. Subsequently central venous pressure increased (21 to 62 mmHg) and a distended abdomen was detected. Intra-abdominal pressure was markedly increased (26 to 40mmHg). Although the venous cannulae were repositioned multiple times no improvement in the patients` condition and the hemodynamic impairment were made. Additionally in one patient an abdominal ultrasound demonstrated subtotal obstruction of the hepatic aspect of the inferior vena cava. So the decision for immediate decompressive laparotomy was made. Median laparotomy was performed and no ischemia, but intestinal swelling and a relevant amount of ascites were found. The chest was left open in three and the small bowl was placed in a gastrochiasis bag in all patients.

One patient died immediately after surgery due to a hypocalcaemia related asystole. The three survivors had an uncomplicated course with relocation of the abdominal content within five days.

Abdominal compartment syndrome is a rare and dangerous complication, which can be treated successfully by early decompressive laparotomy.

The mechanism leading to secondary abdominal compartment syndrome in the setting of elective infant cardiac surgery utilizing extracorporeal circulation is poorly understood. Initially we believed that the event may be due to the cannula type or the cannulation technique in the IVC, however two different surgeons were involved and in one case a single right atrial cannula was used.

We hypothesis that striving for high-flow CPB in small infants and maintance of this flow by addition of excessive amount of fluids may result in liver expansion and congestion and thereby compression of the inferior vena cava, with subsequent further impairment of venous return.

Learning from experience (Congenital): Room Jura **(14:00-15:00)**

Eternal ECMO: The challenge of prolonged post-cardiotomy extracorporeal membrane oxygenation

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Extracorporeal Membrane Oxygenation (ECMO) is the most readily available and widely used mode of postoperative mechanical cardiopulmonary support in pediatric cardiac surgery. Yet, the relatively prompt development of major complications limits its usefulness to short-term support, so it is typically used as a bridge to either recovery or to initiation of ventricular assist device support.

However, over the last few years, many improvements in technology and techniques, increased awareness of potential complications, meticulous preventive and supportive individualized patient management entailing development of specific patient – care protocols in the context of a specialized team approach have resulted in increased duration of effective patient support, thereby maximizing the patient's chances of survival.

Important technological developments have included:

a) The introduction of coated circuits including oxygenators, heat exhangers, tubing and cannulas ("tip-to-tip" coating) in order to minimize the systemic inflammatory response and the need for high levels of anticoagulation.
b) The shift from occlusive roller pumps to centrifugal pumps and, more recently, to axial magnetically levitated pumps that have minimized hemolysis and its serious complications, especially kidney failure.
c) The shift from flat sheet silicone membrane to hollow fiber oxygenators.
d) The development of in-line monitoring equipment for oxygen saturation, blood gases and electrolyte measurement, and for assessment of cerebral and peripheral organ oxygenation.
e) The development of methodology to easily monitor heparin levels and platelet function as well as to monitor coagulation factors has been invaluable for optimal anticoagulation management.

Important developments in patient management have included:

a) Increased awareness of the need to institute ECMO early and promptly, and the organization of rapid ECMO deployment systems and teams.
b) The development of strategies to monitor for, diagnose early and adequately treat subtle but lethally deficient tissue malperfusion.
c) Application of strategies to minimize bleeding and its complications.
d) Increased awareness of the need to aggressively look for and correct undiagnosed or residual lesions that may preclude recovery. Transesophageal echocardiography as well as other major diagnostic procedures, both noninvasive (computerized tomography) and invasive (cardiac catheterization and angiography) can be performed safely under ECMO. Reparative cardiac operations can also be performed under ECMO support, utilizing a period of conversion to conventional cardiopulmonary bypass if "open" procedures are needed.
e) Aggressive management of tissue edema (peritoneal dialysis and in-line ultrafiltration), which can seriously impair organ function.
f) Efficient control of renal failure (employment of continuous hemodialysis)
g) The development of strict patient infection prevention and treatment protocols.
h) Intensive nutritional support, enteral whenever possible, is key during prolonged support.
i) Specific protocols but also an individualized approach to determine the degree of recovery and achieve safe weaning at the earliest possible time.

All aspects of care of the ECMO patient require a coordinated multidisciplinary team approach and dedicated meticulous care by surgeons, cardiologists, intensivists, anesthesiologists, perfusionists, and nurses, while systematic communication with the family regarding the realistic prospects for recovery is essential.


George Sarris

Our presentation pertained the case of an infant with functional single ventricle who required postoperative cardiopulmonary support with ECMO. Although the patient's underlying pathology did not permit ultimate recovery, employment of many of the technological and methodological improvements alluded to above did allow us to provide effective and, by ECMO standards, quite prolonged support (144 days).

ECMO support can be severely taxing to the technical, human and psychological resources of the pediatric cardiac unit, especially if more than one patients are supported simultaneously. A team approach incorporating many of the recent advances outlined above can facilitate facing such challenges efficiently, optimizing chances to support our patients effectively for longer periods of time and to maximize the probability for their survival.