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Transcatheter valvotomy in neonates with pulmonary atresia with intact ventricular septum and duct-dependent pulmonary valve stenosis: who needs complementary intervention?

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Transcatheter pulmonary valve perforation/dilation has improved outcome of neonates with pulmonary atresia-intact ventricular septum (PA-IVS) and duct-dependent pulmonary valve stenosis (PVS). We investigated parameters related to Blalock-Taussig (BT) shunt following successful transcatheter procedure and mid-term outcome.

From 2003 to 2015, 65 consecutive neonates with PA-IVS (n=29) and duct-dependent PVS (n=36) underwent transcatheter pulmonary perforation/dilation and were retrospectively included.

Catheterization (median age 4 days, median weight 2.9 kg) was successful in 56/65 (86%). Among the 6 remaining patients, 1 died (PA perforation) and 5 underwent surgical valvotomy associated with a BT shunt in 3 (1 death). After successful catheterization, 9 patients required a complementary BT shunt. BT shunt was needed more frequently in patients with smaller tricuspid annulus (9.9 vs 12.8mm; p=0.033; Z-score: –1.9 vs –0.8, p=0.03); smaller pulmonary annulus (5.4 vs 7.1mm; p=0.003; Z-score: –2.5 vs –1.5, p=0.03) and with bipartite right ventricle (44.4% vs 14.0%; p=0.05). Shunt intervention was related to longer mechanical ventilation (t=0.0002), in hospital stay (p=0.04) and higher severe morbidity (12.6% vs 55.6%; p=0.01) but not mortality. Global severe morbidity (18.7%) included e.g. necrotizing enterocolitis and septic shock. Global early mortality was 11% (n=7/65). Among the 58 survivors, 56 patients (96.5%) achieved a biventricular circulation after a median follow-up of 1 year [0.2-3.3], including 7 with a previous BT shunt. Two patients were repaired on a one and a half ventricle strategy following BT shunt.

Transcatheter valvotomy for PVS or PA-IVS is efficient to achieve biventricular repair in most suitable candidates. However, in neonates with a small tricuspid valve and/or pulmonary valve annulus, a systemic to pulmonary shunt can be necessary. Even if long-term mortality and morbidity remains an important issue, an excellent mid-term outcome is expected.

Conflict of interest The authors have not transmitted any conflicts of interest.

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Outcomes after protein-losing enteropathy in univentricular hearts: A multicenter study

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Protein-losing enteropathy (PLE) is a rare but severe complication after Fontan surgery in patients with univentricular hearts (UVH), with compromised outcomes since mortality is high and treatment efficiency limited.

Methods A retrospective observational study was carried out in 16 Pediatric Cardiology Centres in France, including all UVH patients diagnosed with PLE after Fontan surgery (study period from 1988 to 2014).

Results PLE was diagnosed in 35 patients at a median age of 9.7 years and a median delay after Fontan surgery of 3.6 years. Cardiac catheterization at diagnosis revealed a biventricular dysfunction of the Fontan circulation in 63%. Treatment modalities included medical treatment alone (MT) in 46% (n=16/35) or combined treatment (CT) i.e. interventional or/surgical therapy associated to medical treatment) in the other 54% (n=19/35). Treatment was considered to be efficient in case of normalization of albumin level (>30g/l). MT led to complete recovery in 13% (n=2/16), to transient improvement in 31% (n=5/16) and to no improvement in 56% of patients (n=9/16). Two MT patients (13%) were transplanted: one recovered, one died. CT led to 21% recoveries (n=4/19), 37% transient improvements (n=7/19) and to no improvement in 42% patients (n=8/19). 37% (n=7/19) in CT group died.16% (n=3/19) underwent heart transplantation but all of 3 subsequently died. Five and 10 years survival were 89.7% (CI95%±1.3%) and 74.9% (CI95%±2.1%) respectively (median follow-up: 4.5 years [0.5-21.7]). Univariate Cox analysis did not reveal any risk factor for “death” nor for “death and transplantation”.

Conclusions Despite decreased mortality, PLE remains a significant burden after Fontan surgery since complete recovery is rare and treatment modalities are unsatisfying. Heart transplantation as ultimate therapeutic option carries a high risk. Further studies are needed to develop innovative treatment strategies and improve outcomes.

Conflict of interest The authors have not transmitted any conflicts of interest.

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Does acute kidney injury increase respiratory morbidity in post-operative course in tetralogy of Fallot?

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Objective Acute Kidney Injury (AKI) is a frequent complication after a pediatric cardiac surgery, especially in neonate and in pre-load-dependent patients, as Tetralogy of Fallot (TOF). It has been suggested that early extra-renal replacement could provide a benefit to neonate. In Pediatric Cardiac Intensive Care Units, an aggressive protocol of fluid management has to be defined. Our goal in patients operated of TOF was to study if AKI was related to surgical parameters and influenced respiratory morbidity.

Methods 60 patients, under 12 months, who underwent elective surgery for TOF, were studied retrospectively. Based on validated pRIFLE criteria they were separated in 2 groups: the first with a presentation AKI (AKI+)+ and the second no (AKI–). The median duration of mechanical ventilation, the median length of by-pass and aortic clamping, the pulmonary valve preservation were significantly longer (3 days versus 1 day, p<0.05). The duration of mechanical ventilation was significantly longer (3 days versus 1 day, p≤0.05) in the AKI+ group.

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