Given the morbidity associated with paradoxical systemic embolization, the site of which is most frequently cerebral, paradoxical embolus in transit may be considered a strong indication for surgery. This requires confirmation by larger studies, ideally in a prospective fashion, which is unlikely to be performed owing to several logistical obstacles, including the rarity of the disease. Until then, the treating clinician must use judgment combined with data from case reports and “meta-analyses” of these to make the best individualized decision for each patient.

References


Successful Palliation of a Child With Left Ventricular Noncompaction Cardiomyopathy and Tricuspid Atresia to Fontan Procedure

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Fig 3. Transesophageal echocardiography (midesophageal four-chamber view) demonstrating inferior vena cava tumor thrombus extending into the right atrium and across a patent foramen ovale into the left atrium.

Comment

Pulmonary embolectomy is usually reserved for significant hemodynamic instability or contraindications to thrombolysis. Other less common scenarios lending themselves to surgical therapy include right-side heart thrombus, namely, “clot in transit,” and thrombus straddling a PFO, or “paradoxical embolus in transit” with impending arterial thromboembolism. In these scenarios, the risks of surgery are outweighed not only by the risk of bleeding with thrombolysis, but also by the occasionally described risk of thrombolysis-induced clot fragmentation and embolization [7].

The literature contains several reports of paradoxical emboli in transit, with no clear consensus on management. Few analyses of the existing reports have been published. Myers and colleagues [7] analyzed 174 patients from 154 studies of impending paradoxical embolism. Surgical thromboembolectomy demonstrated a significant reduction of the composite of mortality and systemic embolism compared with anticoagulation therapy or thrombolysis [7]. In a separate, similar analysis by Fauveau and associates [6], 36 of 88 patients with thrombus straddling a PFO presented with systemic embolism before treatment, with the most common site being cerebral in 26 patients. These findings support the surgical treatment of paradoxical embolus in transit. Furthermore, PFO closure is a potential benefit of surgical embolectomy in preventing recurrent paradoxical embolism.

We present 3 patients with paradoxical embolus in transit, 2 with a preoperative diagnosis as part of the workup for massive or submassive pulmonary embolus and 1 who likely embolized intraoperatively during caval and right atrial tumor thrombectomy. With advances in perioperative care, pulmonary embolectomy can be performed with excellent survival rates, with adverse outcomes mainly related to preoperative hemodynamic instability [8]. In the current report, no patient had systemic embolization, and all had unremarkable postoperative courses and were alive and well several months postoperatively.
A newborn baby girl was diagnosed with tricuspid atresia, ventricular septal defect, normally related great vessels, and hypoplastic right ventricle with no pulmonary stenosis. There was extensive left ventricular non-compaction cardiomyopathy. The left ventricular ejection fraction was 58%. She underwent placement of a pulmonary arterial band at 4 weeks of age, a bilateral bidirectional Glenn at 10 months of age, and fenestrated extracardiac total cavopulmonary connection at 3 years of age. The presence of left ventricular noncompaction in a patient with univentricular circulation does not necessarily circumvent successful cavopulmonary palliation, when left ventricular function is relatively preserved, as in this patient.


Left ventricular noncompaction cardiomyopathy has become widely recognized over the last decade as a leading cause of cardiomyopathy [1]. Studies report it accounting for as much as 9% of cardiomyopathy cases [2]. The occurrence of left ventricular noncompaction cardiomyopathy in association with congenital cardiac defects, including univentricular hearts, has also been well described [3]. We describe the first report of a child with tricuspid atresia and left ventricular noncompaction cardiomyopathy who progressed to Fontan procedure.

A baby girl was born at term weighing 3.2 kg. She was noted to be cyanosed on day 2 of life with oxygen saturations of 78%. Clinical examination revealed normal pulses, preductal and postductal saturation of 78%, a normal first and single second heart sound. There were no audible murmurs. There was a 1 cm hepar. Electrocardiogram demonstrated right atrial enlargement and left axis deviation.

Echocardiogram demonstrated situs solitus with levo-cardia. There was tricuspid atresia. The atrial septal defect was nonrestrictive with right to left shunting. There was a nonrestrictive ventricular septal defect. There were normally related great vessels with no significant pulmonary stenosis. The aortic arch was left-sided with a small patent ductus arteriosus with low velocity left to right shunt. The pulmonary venous drainage was to the left atrium. There were bilateral superior caval veins with a left superior vena cava to coronary sinus connection with no bridging vein. The right ventricle was hypoplastic. The left ventricle was globular with extensive trabeculations and deep recessions. The noncompaction to compaction ratio exceeded 2:1. The transmitral inflow pattern demonstrated a restrictive filling pattern. The left ventricular ejection fraction was 58%. Metabolic and genetic testing was normal. A muscle biopsy was normal.

At 4 weeks of age, she underwent placement of a pulmonary arterial band through a left-sided thoractomy. Postoperative echocardiogram demonstrated a well-placed band with peak gradient 55/60 mm Hg across the band. At 10 months of age, she underwent a bilateral bidirectional Glenn shunt and atrial septectomy. She had a postoperative chylothorax but the procedure was otherwise uncomplicated. She was treated with diuretics, digoxin, aspirin, and captopril.

Cardiac catheterization was undertaken at 3 years of age. The pulmonary arterial band was in good position. There was a nonrestrictive atrial communication. The bidirectional Glenn shunts were widely patent. The pulmonary arterial pressure was 16/11 mm Hg (mean 13 mm Hg), left atrial pressure was 13/10 mm Hg, left ventricular end-diastolic pressure was 11 mm Hg, and the femoral arterial pressure was 79/36 mm Hg (mean 49 mm Hg). Left ventricular angiography outlined the extensive trabeculations and recesses (Fig 1). There was good left ventricular systolic function. An extracardiac total cavopulmonary connection was created with a 20-mm Gore-Tex (W. L. Gore, Flagstaff, AZ) vascular graft, and a 5-mm fenestration was fashioned from the lateral wall of the right atrium to the graft. The pulmonary valve was oversewn to remove risk of thromboembolism. There were no postoperative complications. She was extubated on day 2, chest drains removed day 7, and discharged home 2 weeks after surgery. Four years after Fontan procedure, the fenestration has spontaneously closed, the child is asymptomatic, has excellent exercise capacity, and is free of arrhythmia and protein-losing enteropathy.

Comment

The case details the first report of a child with left ventricular noncompaction and tricuspid atresia progressing to Fontan procedure. Although previous reports have highlighted the association of single ventricle patients and left ventricular noncompaction, our report highlights

Fig 1. Angiographic evidence of extensive left ventricular noncompaction. Note numerous trabeculations and recesses.
that surgical palliation is also possible in this group of patients [1, 3, 4]. Preoperative catheterization demonstrated favorable hemodynamics with a mean pulmonary arterial pressure 13 mm Hg, good-sized branch pulmonary arteries, and good left ventricular systolic function, although the left ventricular end-diastolic pressure was mildly elevated at 11 mm Hg. It was elected to fenestrate the extracardiac conduit to minimize the risk of low cardiac output after the Fontan procedure.

Our patient had well-preserved left ventricular systolic function, which prompted us to progress her through the various univentricular palliations. In fact, the Glenn procedure may have benefited the left ventricular function by off-loading the single ventricle. Clearly, an alternative strategy would be required were the left ventricular systolic or diastolic function significantly compromised or in the presence of significant arrhythmia [5].

In conclusion, patients with left ventricular noncompaction cardiomyopathy and tricuspid atresia/hypoplastic right ventricle may tolerate univentricular palliations, including Glenn and Fontan procedures, when the left ventricular function is relatively preserved.

Tricuspid atresia and common truncus arteriosus are rare forms of congenital heart disease; the coexistence of both anomalies is therefore an extremely uncommon event. Without treatment, early mortality is the natural course so diagnostic and therapeutic management must be performed without delay. We report a case of a newborn with a postnatal diagnosis of coexistent tricuspid atresia and common arterial trunk in whom successful palliation was performed using a staged surgical approach.

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A female infant twin was born at 41 weeks’ gestation, weighing 1,746 g. There was no antenatal cardiac diagnosis for either of the babies. Her twin had a postnatal diagnosis of coarctation and ventricular septal defect (VSD) on day 6 of life. Our patient was initially clinically well, self-ventilating in air. On day 9 she exhibited an acute deterioration, with cardiopulmonary decompensation, respiratory distress, cyanosis, and severe metabolic acidosis. She required resuscitation with inotropic and ventilatory support. A transthoracic echocardiogram performed at the time showed TA with a hypoplastic right ventricle, a CTA type 1.5/2 with mild truncal valve regurgitation, an unrestrictive interatrial communication, and good-sized branch pulmonary arteries.

On day 12, with a weight of 2,200 g, she underwent her first operation. Through a medium sternotomy, anatomic inspection confirmed CTA with the two pulmonary artery branches arising separately but with very proximate origins posteriorly from the aorta. Using cardiopulmonary bypass with intermittent cold blood cardioplegia, the truncus was opened and a patch of Gore-Tex (W.L. Gore & Assoc, Flagstaff, AZ) was used to separate the aorta from the pulmonary arteries. After this, a Gore-Tex 4-mm graft was interposed between the right ventricle and the pulmonary outflow tract, thereby creating a Sano shunt (Fig 1). An atrial septectomy was then performed. The patient was weaned off cardiopulmonary bypass with good hemodynamics, and an epicardial echocardiography showed a satisfactory repair with good biventricular function.

Tricuspid Atresia With Truncus Arteriosus: Successful Surgical Treatment
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