Increasing cyanosis early after cavopulmonary connection caused by abnormal systemic venous channels

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Abstract

Objective—To show that abnormal systemic venous channels in patients who undergo cavopulmonary anastomoses can become manifest and haemodynamically important only after surgery despite detailed preoperative investigation.

Design—Descriptive study of patients fulfilling the above criteria selected from hospital records over the past three years.

Setting—A tertiary referral centre.

Patients—Of the three cases identified, two were isomeric, one with left atrial isomerism and hemiazygous continuation of the inferior vena cava who underwent bilateral bidirectional Glenn anastomoses and one with right isomerism who underwent total cavopulmonary anastomosis. Case 3 had absent left atriovenous connection with a hypoplastic left lung and underwent a classic right Glenn procedure. All three cases presented with progressive cyanosis in the early postoperative period.

Interventions and results—Postoperative angiography in case 1 showed a remnant of a left inferior vena cava draining to the atrium to have become grossly dilated causing cyanosis, which resolved after redirection of this vessel and of the hepatic veins into the right pulmonary artery with an intra-atrial baffle. Cyanosis in case 2 was caused by intra-hepatic shunting to a hepatic vein draining to the left of the intra-atrial baffle. The diagnosis was made at necropsy, being overlooked on postoperative angiography. Repeat angiography in case 3 showed progressive dilatation of a small left superior vena cava to coronary sinus. Test occlusion with a view to embolisation revealed hitherto an underdramatised hemiazygous continuation of inferior caval to brachiocephalic vein. The patient underwent surgical ligation of these two venous channels.

Conclusions—Despite appropriate investigation some “abnormal” venous pathways manifest themselves, dilate, and become haemodynamically important only after surgical cavopulmonary anastomoses. In the presence of early postoperative cyanosis “new” systemic venous collateral channels should be considered as a possible cause, which may require reintervention.

Keywords: Congenital heart defects, surgery, cyanosis.

Since the first report by Fontan and Baudet in 1971 of right heart bypass for tricuspid atresia, this operation has been modified and its indications extended to more complex lesions. Superior cavopulmonary anastomosis, as a modification of the Fontan procedure, may be employed as definitive palliation, or more usually as interim palliation before completion of total cavopulmonary anastomosis. The latter requires an intra-atrial baffle to direct inferior vena caval and hepatic flow to the pulmonary arteries. The coexistence, development, or dilatation of abnormal systemic venous channels, commonly but not exclusively seen with atrial isomerism, may pose additional problems at, or after, repair. In this study we describe three cases in which these channels should be considered in a patient with residual or progressive cyanosis after such procedures.

Case reports

CASE 1
A 7 year old girl with double outlet right ventricle, subpulmonary ventricular septal defect and severe pulmonary stenosis was referred for further management. She was initially well after a Waterston shunt in Russia, but developed increasing cyanosis with reduced exercise tolerance. Echocardiography showed left atrial isomerism, biventricular atriovenous connexion via two atriovenous valves with right hand topology, and double outlet right ventricle with a very large subpulmonary ventricular septal defect. There were bilateral superior vena cavae with hemiazygous continuation to the left superior vena cava. Finally, there was severe subpulmonary stenosis with the pulmonary artery situated posteriorly and to the right of the aorta. Cardiac catheterisation and angiography confirmed the basic anatomical diagnosis. There was no brachiocephalic vein and bilateral or symmetric pulmonary venous drainage. The Waterston anastomosis was patent and the pulmonary arteries were of good size with a mean pulmonary artery pressure of 9 mm Hg. Biventricular repair was considered to carry a high risk and so a bilateral bidirectional Glenn anastomosis with takedown of the Waterston anastomosis and transection of the main pulmonary artery was performed. The hepatic veins were left to drain directly to the right atrium. The operation was uneventful and the patient returned from the operating room in sinus rhythm and with an oxygen saturation of 75%.
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Figure 1 Unobstructed left Glenn anastomosis. Note the hemiazygos continuation to the left superior vena cava. LPA, left pulmonary artery.

level of 88–89%. She remained well but gradually developed increasing cyanosis. Her oxygen saturation level in oxygen decreased to 65% on the 6th postoperative day and she underwent repeat angiography, which showed unobstructed Glenn anastomoses on both right and left sides (fig 1). Injection into the right femoral vein showed a left sided vein (fig 2(A)) receiving venous return from the gut and kidneys and draining with the hepatic veins into the atrial chambers (fig 2(B)). This was thought to be a remnant of a left inferior vena cava, not detectable on preoperative screening, and clearly contributing to progressive cyanosis as dilatation occurred after operation. The patient was taken back to theatre where she had an intra-atrial Gore-tex baffle redirecting the left abdominal vein with the hepatic veins into the right pulmonary artery via the right superior vena cava. The patient’s oxygen saturation level rose to 97% with a left superior vena caval mean pressure of 15 mm Hg and systemic systolic blood pressure of 80 mm Hg. She made an uneventful recovery and was discharged home 1 week later with oxygen saturation levels in air of > 95%.

CASE 2

A 2 year old girl with severe cyanosis and breathlessness on minimal exercise was referred from Yugoslavia for assessment and surgery. Echocardiography showed right atrial isomerism with univentricular atrioventricular connexion to a dominant right ventricle via a common atrioventricular valve. There was double outlet right ventricle with severe subvalvular pulmonary stenosis. She had bilateral superior vena cavae with no bridging innominate vein. The inferior vena cava drained to the right side and all the pulmonary veins to the left side of a common atrium. The pulmonary arteries were confluent and of good size. Cardiac catheterisation confirmed the anatomical diagnosis. There was minimal atrioventricular valve regurgitation and the mean pulmonary artery pressure was 12 mm Hg. She underwent total cavopulmonary anastomosis with bilateral bidirectional Glenn shunts, and a fenestrated (4 mm) intra-atrial impa baffle rerouting the inferior vena cava to the right pulmonary artery opposite to the right Glenn anastomosis. The main pulmonary artery was trans-sected and sutured. She returned to the intensive care unit in sinus rhythm and with oxygen saturation levels of 70–75%. She was extubated the following day. Between the 2nd and 3rd postoperative days the patient developed increasing cyanosis with oxygen saturation levels of 60–64%. She had repeat cardiac catheterisation which showed unobstructed flow through all the anastomoses. Balloon closure of the fenestration raised the systemic oxygen saturation transiently to 80% but this returned to 50–60% after a few minutes. The procedure was abandoned because the patient went into junctional rhythm and became
unwell. Her condition deteriorated rapidly because of junctional tachycardia that was unresponsive to conventional treatment. She developed extreme cyanosis and acidosis and died the following day. Postmortem examination showed a "classical example" of right isomerism, with symmetrical features including bronchi, appendages, and caval veins. The surgical anastomoses were intact and there was a large hepatic vein draining to the left side and posteriorly into the common atrium (fig 3). Although overlooked at the time, on careful review of the injection into the inferior vena cava there was late preferential filling of the atrium due to intrahepatic shunting to a hepatic vein draining directly to the left of the intra-atrial baffle (fig 4).

CASE 3

A 20 month old boy with situs solitus, absent left atrioventricular connexion, and hypoplastic left lung was referred for surgery because of severe cyanosis. Echocardiography showed usual atrial arrangement, concordant atrioventricular connexions, and double outlet right ventricle with severe subpulmonary stenosis. The systemic and pulmonary venous returns were thought to be normal. There was an imperforate left atrioventricular valve, a mildly restrictive atrial septal defect, and the left ventricle was hypoplastic. Cardiac catheterisation and angiography showed occlusion of the left pulmonary artery at the insertion of the duct with extreme hypoplasia of the distal left pulmonary artery and lung. A previous left modified Blalock-Taussig shunt was also occluded. There were no significant systemic to pulmonary collateral arteries to the left lung. A right classical Glenn anastomosis and atrial septectomy were performed. It was not possible to establish continuity between the main and left pulmonary artery because of the long segment of occlusion and severe hypoplasia of branch vessels. The patient returned from the theatre in sinus rhythm with oxygen saturation levels of 80–84%. Despite continuous systemic heparinisation thrombosis at the junction of brachiocephalic to right superior vena cava was diagnosed when he developed increasing cyanosis (40–50%) on the 4th postoperative day. Angiography performed to demonstrate the thrombus showed a small left superior vena cava draining to the coronary sinus (fig 5). The patient was treated with streptokinase and repeat angiography 1 week later showed complete resolution of the thrombi and the patient’s oxygen saturation level had increased to upper 78%. One week later, although the patient was relatively well, he gradually developed increasing cyanosis with average oxygen saturation levels of 57%. Repeat angiography then showed a more dilated left superior vena cava draining to the right atrium via a coronary sinus. Balloon occlusion of the left superior vena cava (with a
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Discussion

Cavopulmonary anastomosis is increasingly employed for patients with complex congenital heart disease when a biventricular repair is not possible. Superior cavopulmonary anastomosis (bidirectional Glenn) provides excellent interim palliation with low morbidity and mortality. Similarly, total cavopulmonary anastomosis, particularly when fenestrated, has led to more rapid postoperative recovery and appears to have reduced mortality in complex high risk cases. Our report shows that the coexistence and complexity of abnormal systemic venous return commonly but not exclusively seen in isomorphic cases represents an additional challenge to the management of these patients. The result may be an increase of right to left shunting driven by the high venous pressure in the systemic veins. Some of these channels can and should be diagnosed before operation if a careful descriptive approach is pursued by analysing the veins returning to the heart one at a time in a sequential fashion—that is, superior vena cavae, the coronary sinus return, the inferior vena cavae, the azygos system, and the hepatic veins. Any scheme of description must describe the presence or absence of each segment and its connexions. Atrial situs is a key variable in the understanding of venous developement. The range spectrum of abnormalities is relatively limited if there is lateralised situs. In case 3 there was a left superior vena cava to coronary sinus, which is seen in 3% of cases of situs solitus. There was also hemiazygos continuation which is extremely rare in this setting. When the situs is abnormal the connexions of the venous segments are always abnormal, but again to some extent still predictable—that is, absent coronary sinus in almost all cases of right atrial isomerism.

Previous reports refer to both technical difficulties at repair and increased perioperative mortality associated with atrio pulmonary Anastomotic procedures in combination with intra-atrial rerouting of abnormal venous channels. More recent reports of cavopulmonary anastomoses emphasize the need for a detailed preoperative anatomical and physiological diagnosis and an individualised plan for each patient to provide unobstructed venous pathways. Unlike others, our report focuses on severe progressive cyanosis in the immediate postoperative period after cavopulmonary anastomoses despite appropriate preoperative investigation. This is a matter that has not received attention in the literature. All three patients had preoperative echocardiographic and angiographic studies to demonstrate venous anatomy. None of the channels that became apparent after operation were demonstrated before surgery. Indeed, these channels may become anatomically and physiologically important only after surgery when they dilate. It may not be possible to demonstrate these “potential” channels with any imaging modality, although in case 2, in which a single hepatic vein drained separately to the left side of the atrial septum, allowing an intrahepic right to left shunt, this should perhaps be defined by preoperative echocardiography.

Thus, in summary, despite appropriate investigation some abnormal venous pathways will manifest themselves or become haemodynamically significant only after surgery. If this situation occurs then these venous channels can form new bypass circuits at variable levels with right to left shunting, which may lead to increasing cyanosis. Awareness of this complication should alert physicians dealing with progressive postoperative cyanosis after cavopulmonary anastomosis to reinvestigate their patient’s systemic venous channels, as some may require reoperation.

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