Case report - Congenital

Late desaturation due to collateral veins 10 years after total cavopulmonary shunt in left atrial isomerism: surgical closure

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Abstract

The development of systemic collateral veins after palliative surgery in children with univentricular circulation is a common complication, however, manifestation as late as 10 years postoperatively is rare. Massive systemic to hepatic venous collaterals developed in a 14-year-old girl with univentricular heart, situs inversus atriovisceralis and hemiazygos continuity to the left-sided superior vena cava, 10 years after Kawashima operation. The resulting azygoportal shunt had led to a progressive systemic desaturation and reduction in ventricular function. Interventional occlusion was supposed to be risky for renal failure due to potential closure of the renal vein so that surgical closure was performed. The saturation persistently increased from 65% to more than 85% postoperatively.© 2008 Published by European Association for Cardio-Thoracic Surgery. All rights reserved.

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1. Introduction

Patients with univentricular circulation and azygos or hemiazygos connection of the infrahepatic inferior vena cava (IVC) who had been palliated with cavopulmonary shunt may later develop collateral veins leading to systemic desaturation [1]. Only closure of these collaterals or Fontan-type surgery may contribute to an improved saturation.

2. Case report

We present the case of a 14-year-old girl with complex congenital heart disease consisting of double inlet right ventricle, D-transposition of the great arteries, pulmonary atresia, hemiazygos continuity (azygos in situs inversus) for the IVC and situs inversus atriovisceralis. With Kawashima operation (total cavopulmonary shunt) performed 10 years before the systemic venous return, except for hepatic and coronary venous flow, directly drained into the pulmonary artery.

The girl had grown up well and besides dyspnea at extensive exertion no other disorder existed. In the annual follow-up the systemic saturation had been between 86 and 92% for years. Recently, however, over the last six months progressive cyanosis with impaired exercise tolerance developed. Echocardiography revealed reduced ventricular function. In the graded exercise test electrocardiographic response was positive, the saturation decreasing to as low as 65% at rest. Angiography detected multiple venous collaterals originating from the right renal vein with resulting azygoportal shunt (Fig. 1a). Interventional occlusion was considered to be risky particularly in this case for potential thrombosis of the right renal vein. Therefore, we favoured surgical occlusion.

The situs inversus atriovisceralis added to complexity. After identification of the 10 mm measuring right renal vein we began to ligate and partially excise all venous collaterals originating from the renal vein. The saturation had improved significantly from 65% to levels around 85% under ventilation (FiO 0.4). In the following days renal function, serum creatinine and urea levels remained normal and saturation was found to be between 85 and 95%. In the control angiography on the 3rd POD no major venous collaterals could be detected. Two smaller left over collaterals were immediately treated with percutaneous coil embolisation (Fig. 1b). The girl was discharged home on the 13th POD. During the current follow-up the patient presents as a young woman with 54 kg weight at a height of 172 cm. The saturation remained at levels between 85 and 95%. The current angiography (Fig. 2a), after now 2.5 years postoperatively, did not reveal a recurrence of the collateral veins and there is still the lack of formation of pulmonary artero-venous fistulae (Fig. 2b). The patient has a satisfying ventricular function with an end diastolic pressure of 9 mmHg.

3. Discussion

The creation of a total cavopulmonary shunt provides a sufficient palliation for children with interrupted IVC and
complex congenital heart disease. In most of these children with univentricular circulation after Glenn or Fontan operations, the increased central venous pressure may induce recanalization of embryologically preformed and obliterated vessels [2–4]. Finally, such venous collaterals can result in systemic desaturation and reduction in ventricular function [1, 5]. As presented in our case, venous collaterals developed late resulting in ayzygoperitral shunt causing further desaturation. The prevalence of such collaterals is considered as a common complication after cavopulmonary connection and varies between 20.2% [1] to 31% [3] of the patients. A late manifestation as in our patient, however, is extremely rare [4].

While Fontan completion is one choice of treatment, the other therapeutic option to eliminate such collaterals consists of percutaneous coil embolisation. A smaller number of patients underwent surgical ligation of collaterals. As presented in our case the venous collaterals originated directly from the right renal vein. Percutaneous coil embolisation...
isation would have led to occlusion by thrombosis of these collaterals, but due to a direct connection to the right renal consecutively thrombosis of the renal vein would be triggered. Uneventful recovery and the current satisfying development of the patient with persisting improved saturation justifies our chosen therapy regimen. Furthermore, the findings of the current angiography, with the lack of recurrence of the collateral veins, now 2.5 years postoperatively, is promising.

Fontan completion was not opted for in this patient as the degree of desaturation had led already to impairment of left ventricular function. Although this is known to reverse after improvement of saturation, the time course of improvement may not yet be beneficial for the early postoperative state after Fontan completion.

In conclusion, non-cardiac surgery as the favoured therapeutic choice for occlusion of such massive venous collaterals may be beneficial in selected patients with impaired ventricular function and rather complex collateral anatomy, who otherwise were high-risk Fontan patients.

References