Patent ductus venosus causing cyanosis after Fontan operation - successful transcatheter-based flow reduction to induce growth of rarefied intrahepatic portal veins allowing for ultimate interventional closure

Kerst G., Khalil M., Moysich A., Schmidt D., Schranz D.
Pediatric Heart Centre, University Children’s Hospital, Giessen, Germany

Background:
Pulmonary arteriovenous fistulas (PAVM) leading to cyanosis after bidirectional superior cavopulmonary anastomosis are considered to be due to hepatic venous blood bypassing the lungs. Here report for the first time that a patent ductus venosus (PDV), a persistent fetal connection between the left portal vein and the inferior cava, should be considered as a cause for PAVM after Fontan operation.

Case report:
PAVM were diagnosed in a 9-year-old boy with episodes of dizziness developing cyanosis after modified Fontan procedure for palliation of pulmonary atresia with intact ventricular septum. During color Doppler examination of hepatic flow a PDV was diagnosed. Via subclavian venous access and a retrograde transcaval route a portal venogram was obtained showing rarefied intrahepatic portal veins (PV). PV pressure increased from 12 to 26 mmHg during temporary PDV balloon occlusion. To avoid PV congestion leading to portal thrombosis, flow reduction of the PDV was achieved by transcatheter placement of a diabolo-shaped covered stent-ensemble consisting of a 5x12 mm Formula renal stent centrally placed on a 16x41 mm covered Advanta V12 stent yielding a gradient of 5 mm Hg between PV and inferior vena cava. Repeat portal venogram 5 months later showed increased vascularity of the PV system and a minor PV pressure increase from 13 to 16 mmHg during temporary PDV balloon occlusion. After ultimate closure of PDV with an 8 mm AVP II, elevated levated blood ammonia levels normalized, episodes of dizziness disappeared and tcSaO2 gradually increased from 80-85 to 85-90% during the following 3 months.

Conclusion:
PDV should be considered as a possible cause for unexplained cyanosis in Fontan patients with PAVM. Transcatheter-based flow reduction of PDV is feasible and appears to induce growth of rarefied intrahepatic portal veins allowing for ultimate interventional closure.