

Successful percutaneous recanalization of the RPA origin after surgical ligation in patient with Ebstein anomaly and Glenn procedure

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Objectives

Absence of a hypothetical hepatic factor passing through the pulmonary circulation is supposed to be responsible for pulmonary arteriovenous fistulas (AVF) development what can lead to severe cyanosis in patients after partial cavo-pulmonary anastomosis (PCPA). Redirection of hepatic blood flow into the pulmonary artery may cause fistulas to close, what significantly improves the clinical state of the patient. In some patients a transcatheter approach for blood redirection of hepatic blood supply into the pulmonary circulation is possible and repeated open-heart surgery can be avoided.

Methods

A 12-year-old girl with Ebstein anomaly, who at the age of 3 years was managed with PCPA and surgical ligation of the right pulmonary artery (RPA) origin, was scheduled for cardiac catheterization due to severe central cyanosis (SaO₂ 80-85%). The right jugular vein and the right femoral vein were cannulated both with 6F sheaths. Angiography showed multiple intrapulmonary fistulas in the right lung. The mean blood pressure RPA: 13 mmHg, LPA: 35 mmHg. Simultaneous injection into SVC and LPA showed the ligated RPA origin with the length of about 5 mm. A radiofrequency wire (Baylis) was advanced through a left Judkins guiding catheter from the jugular access. Repeated energy (up to 15 Watts) applications (under fluoroscopy control) caused slow advancement of the radiofrequency guidewire through the ligated segment. After entering the LPA-lumen the guidewire was snared from the femoral site. A 6F long sheath was introduced from the femoral vein and a 6x16 mm covered LifeStream stent was advanced and implanted into the recanalized segment.

Results

The narrowest segment of the implanted stent was assessed with 2-3 mm what was found optimal due to the increased LPA blood pressure. Control angiography showed no contrast extravasation. After the procedure the clinical state of the patient improved significantly. Three months after stent implantation oxygen saturation was measured with 90%.

Conclusion

Redirection of hepatic blood into the pulmonary artery in patients after PCPA with AVF and severe cyanosis can cause fistulas to close and significantly improve the clinical state of the patient. If possible an interventional approach can be considered in selected patients.