An unusual case of left-sided hemitruncus arteriosus associated with aortic arch hypoplasia

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SOUHRN

Hemitruncus arteriosus patří mezi nejvzácnější vrozené kardiovaskulární malformace. Obvykle postihuje pravou plicní tepnu. Hemitruncus arteriosus se může vyskytnout buď izolovaně, nebo ve spojení s dalšími srdečními vadami, jako jsou defekty septa a Fallotova tetralogie; nicméně se s ní lze pouze zřídkakdy setkat současně s postižením aortálního oblouku.
Popisujeme neobvyklý případ 15denního novorozence s levostranným hemitruncus arteriosus spojeným s hypoplazií aortálního oblouku, koarktací aorty, otevřenou tepennou (Botallovou) dučejí (patent ductus arteriosus) a s kraniofaciálními abnormalitami, u něhož byla provedena chirurgická korekce.

ABSTRACT

Hemitruncus arteriosus is among the rarest congenital cardiovascular defects. Pathology usually involves the right pulmonary artery. It may occur either isolated or associated with other cardiac defects such as septal defects and tetralogy of Fallot; however, it is seldom associated with aortic arch pathologies.
We represent an unusual case of a 15-day-old neonate with left-sided hemitruncus arteriosus associated with aortic arch hypoplasia, coarctation of the aorta, patent ductus arteriosus, and craniofacial abnormalities, who underwent surgical correction.

Background

Hemitruncus arteriosus is among the rarest congenital cardiovascular pathologies, in which a right or left pulmonary artery (PA) emerges directly from the aorta. It accounts for less than 0.1% of all congenital heart diseases. In 1868, Fraentzel was the first to describe this anomaly.[1]

The anomaly may occur as an isolated pathology or in conjunction with other disorders, most frequently associated with patent ductus arteriosus (PDA), less common with ventricular septal defect (VSD), atrial septal defect (ASD), and tetralogy of Fallot (TOF). Association with aortic coarctation and aortic arch hypoplasia is extremely rare.[2,3]

In this case report, we discussed the management of a patient with hemitruncus arteriosus with PDA, aortic arch hypoplasia, coartaction of the aorta, and craniofacial abnormalities.

Case report

A 15-day-old female neonate with a weight of 3 kg and unilateral ear hypoplasia and anophthalmia was admitted to the neonatal care unit due to cyanosis, dyspnea, poor feeding, and fatigue. Examination revealed tachypnea (30 breaths/minute), tachycardia (196 beats/minute), and bilateral pulmonary rales. The oxygen saturation of the neonate was 90%. A chest X-ray indi-

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cated bilateral pulmonary opacities and a mesocardic heart. The echocardiography revealed meso-dextrocardia with atrioventricular and ventriculoarterial concordance. The aorta was located posteriorly and emerged from the left ventricle, measuring 7 mm in diameter. The proximal aortic arch diameter was 4 mm, and the diameter of the distal aortic arch was 3.5 mm. There was a large ductus arteriosus, in which the direction of the shunt was left to right. The main pulmonary artery diameter was 17 mm, and the right pulmonary artery diameter was 6 mm. The diameter of the left pulmonary artery was 4 mm but did not originate from the main trunk but from the anterior aspect of the ascending aorta, 3 mm above the sinotubular junction. The echocardiography findings were confirmed with cardiac catheterization and computerized tomography of the chest, clearly indicating the hemitruncus arteriosus and aortic arch pathologies (**Figs 1–4**).

After explaining the pathology, risks, and benefits of treatment options, we decided on surgical correction with the family's consent. The family also consented to the academic use of the patient's materials.

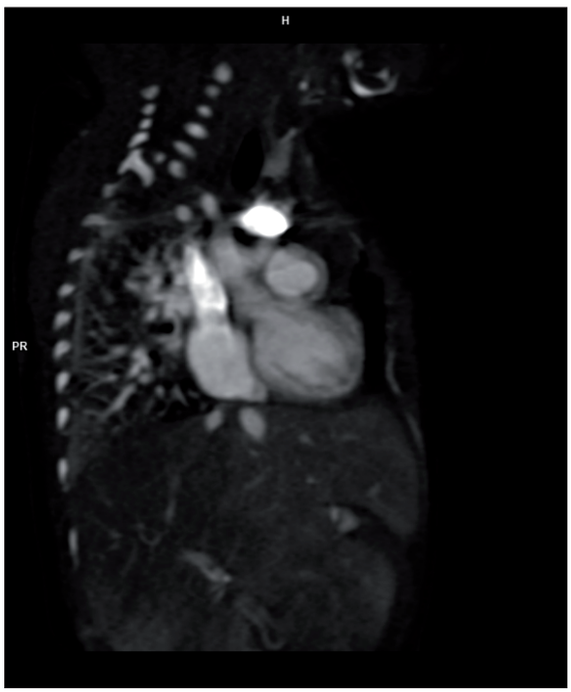

Fig. 1 – Computerized tomography angiography showing the left pulmonary artery originating from the ascending aorta.

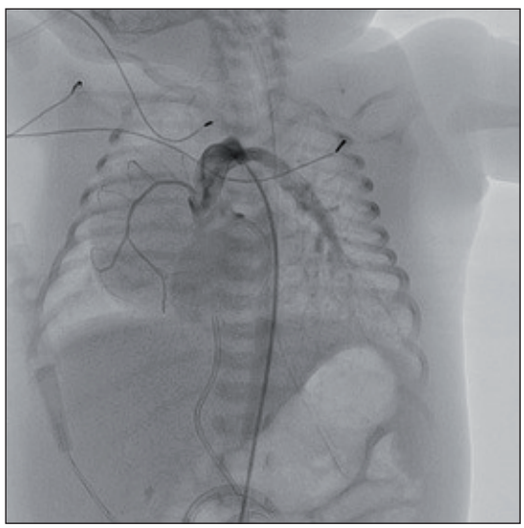

Fig. 2 – Cardiac catheterization showing the left pulmonary artery originating from the ascending aorta.

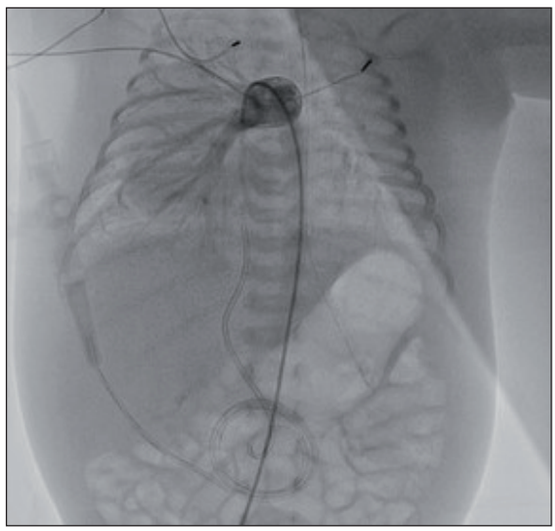

Fig. 3 – Cardiac catheterization showing enlarged main pulmonary artery and course of the right pulmonary artery.

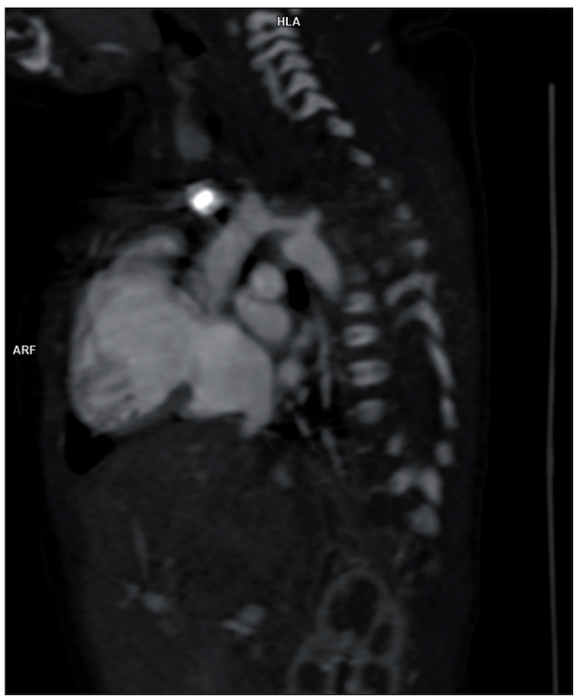

Fig. 4 – Computerized tomography angiography showing the hypoplastic arch and aortic coarctation.

Surgical procedure

A median sternotomy was performed. The heart was dextro-mesocardic. The pericardium was opened and harvested to be used for the aortic arch reconstruction. The ascending aorta gave rise to the left pulmonary artery, which then crossed the main pulmonary artery. The aortic arch was hypoplastic. A 3.5-mm PTFE graft was anastomosed to the innominate artery for antegrade cerebral perfusion. All major thoracic vessels and branches of the pulmonary arteries and ductus arteriosus were prepared and looped. The patent ductus arteriosus was ligated. Cardiopulmonary bypass was commenced with a single right atrial venous cannula and innominate artery. The patient was cooled down to 26 °C. The left carotid and subclavian arteries, the left pulmonary artery, and the descending aorta were clamped. Cardiac arrest was provided with antegrade cold blood cardioplegia following an aortic cross-clamp. The left pulmonary artery was divided from the ascending aorta. The lesser curvature of the ascending aorta was incised beyond the ductus arteriosus, and the arch was reconstructed with fresh pericardium. The aortic cross-clamp was released, and the patient was gradually re-warmed. The left pulmonary artery was anastomosed end to side to the main pulmonary artery with an 8-0 polypropylene. Aortic cross-clamp and cardiopulmonary bypass times were 21 and 56 minutes, respectively.

The patient could be weaned off mechanical ventilation the next day and discharged from the hospital after 6 days. She has been followed actively, symptom-free, and with normal growth for more than 4 months postoperatively; however, she suffers from craniofacial abnormalities that require interventions.

Discussion and conclusions

The abnormally arising vessel in hemitruncus arteriosus is usually the right pulmonary artery, which originates from the posterior aspect of the ascending aorta, a few centimeters above the aortic valve.[4,5] Left pulmonary artery involvement, as in our particular case, in hemitruncus arteriosus is less common.[6]

It is usually associated with other congenital cardiovascular pathologies, being the most frequent tetralogy of Fallot (74%), followed by aortic arch hypoplasia (63%), and PDA (16%).[2] The presentation of the patients varies depending on the associated cardiac anomalies; however, generally, they have heart failure due to pulmonary overflow.

Early diagnosis and timely surgical intervention significantly improve the prognosis of children with this anomaly. Without surgical correction, babies face a staggering 70% mortality rate within the first year, with 30% of them dying within the first three months.[7] Early repair is crucial to prevent the development of pulmonary vascular disease. Ideally, corrective surgery should be performed within the first six months of life to reduce the risk of severe pulmonary atherosclerosis.[7]

The aim is to anatomically implant the pulmonary artery branch into the main trunk with or without cardiopulmonary bypass and correct associated malformations. In our case, we repaired the hypoplastic aortic arch and left-sided hemitruncus with antegrade cerebral perfusion and anastomosis of the left pulmonary artery end to side to the main pulmonary artery without any neurologic consequence.

Conflict of interest
The authors declare that they have no conflict of interest.

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None.

Informed consent
Informed consent was obtained from the patient's family regarding the use of the patient's materials for academic purposes.

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