Hemitruncus arteriosus: a rare congenital cardiac anomaly diagnosed and treated during the neonatal period

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Introduction

Hemitruncus arteriosus is an extremely rare congenital heart defect that accounts for less than 0.1% of all congenital cardiac diseases[1]. It is a subtype of truncus arteriosus where one main pulmonary artery branch originates aberrantly from the ascending aorta[1]. If not recognised and treated, it leads to severe pulmonary hypertension, heart failure and death in early infancy. Here we report a Sri Lankan neonate with hemitruncus arteriosus diagnosed and treated during the neonatal period.

Case presentation

A baby girl was born to a 34-year-old primigravida at term with a birth weight of 2.36kg (-2SD to -3SD) following an uncomplicated antenatal period. There was no parental consanguinity, family history of congenital heart defects or medical complications of the mother. Antenatal ultrasound scans were normal. The length and occipitofrontal circumference at birth were 46cm (-1SD to -2SD) and 34cm (median to +1SD) respectively.

The baby did not cry at birth and required resuscitation for one minute with inflation and ventilatory breaths. Apgar scores were six, nine and nine at one, five and ten minutes. She has had peripheral cyanosis; however, her heart rate was above 100 beats per minute. The baby recovered, and after an initial observation period of two hours, she was handed over to the mother to commence breastfeeding.

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At 24 hours of age, she developed respiratory distress with a respiratory rate of 70 breaths per minute and intercostal recessions. There was no grunting. Vesicular breath sounds were heard bilaterally without any added sounds. Her pulse rate was 130 beats per minute with normal volume bilateral femoral pulses. Auscultation revealed a grade 3 pansystolic murmur best heard at the left lower sternal edge. The liver was palpable 3 cm below the right costal margin, and the spleen was not palpable. Pulse oximetry reading revealed an oxygen saturation of 95% on air. The initial chest x-ray done on day 2 of life was normal (Figure 1).

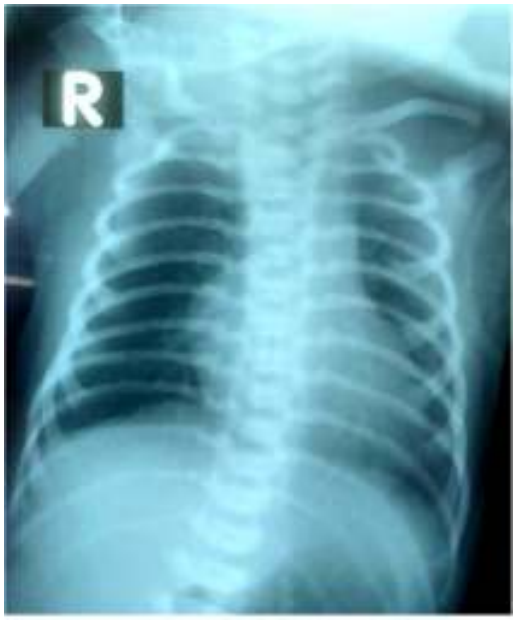

Figure 1: ***Chest x-ray taken on day 2 of life***

An echocardiogram by a paediatric cardiologist on day 2 of life revealed a dilated right atrium and right ventricle and a patent foramen ovale with a bidirectional shunt. The main pulmonary trunk continued as the left pulmonary artery, whereas the right pulmonary artery originated from the aorta 5mm distal to the aortic annulus (Figure 2). There was a left-sided small patent ductus arteriosus and persistent pulmonary hypertension with tricuspid regurgitation with a peak pressure gradient of 97 mmHg. The echocardiography confirmed the diagnosis of hemitruncus arteriosus.

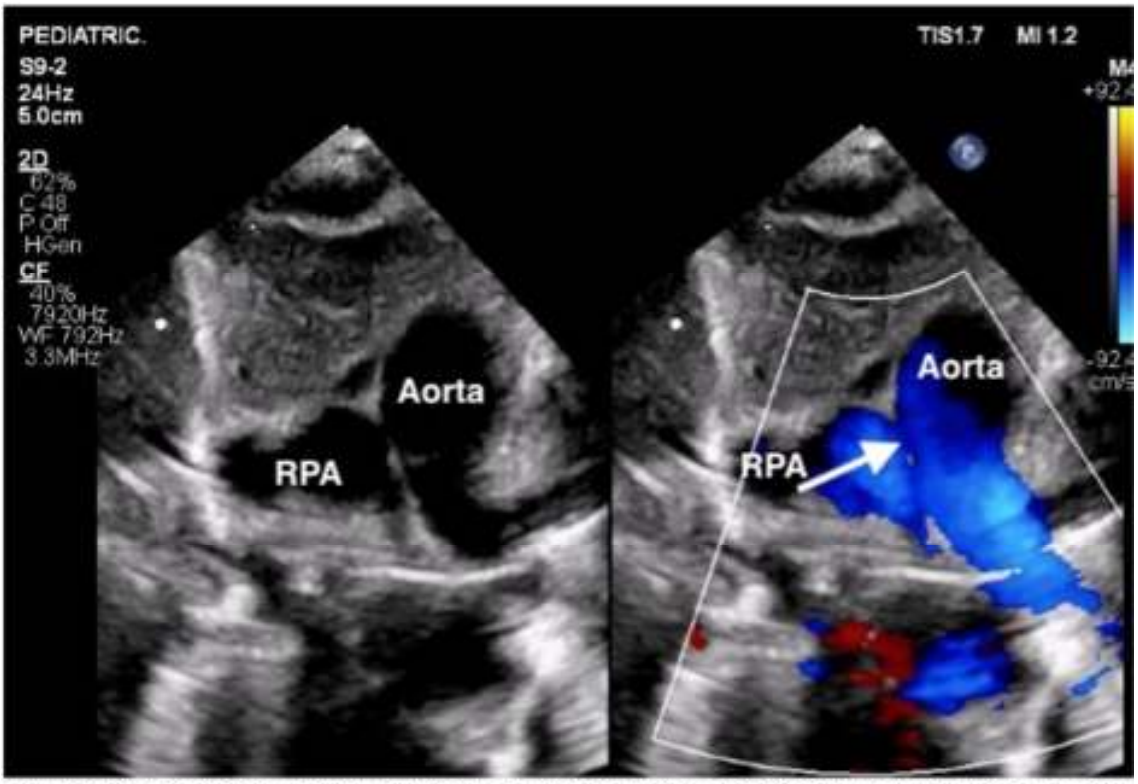

Figure 2: ***Echocardiogram showing right pulmonary artery (RPA) arising from the aorta (white arrow)***

The baby was referred for cardiothoracic surgical care. The corrective surgery with translocation of the right pulmonary artery to the main pulmonary artery and ligation of the patent ductus arteriosus was done on day 28 of life. She had an uneventful recovery. At the three-month review, she was symptom-free and thriving well with a weight of 4.05kg (median to +1SD), length of 50cm (-1SD to -2SD), and occipitofrontal circumference of 39cm (+2SD to +3SD).

Discussion

Hemitruncus arteriosus is a rare congenital heart defect where one main pulmonary artery branch originates aberrantly from the ascending aorta[1,2]. In 75% of cases, the right main pulmonary artery has an anomalous origin, whereas the left pulmonary artery accounts for the remaining 25%[2]. The aetiology is not well understood; however, it is believed to be due to the failure of the development of the sixth aortic arch and the persistence of the fifth aortic arch[3,4].

Severe pulmonary hypertension and heart failure are early complications of hemitruncus arteriosus. This is because one lung receives the entire pulmonary blood volume through the continuation of the pulmonary trunk as one pulmonary artery, while the other lung receives blood at the systolic pressure through the aberrant artery originating from the aorta. Infants usually present with respiratory distress, failure to thrive, generalised oedema, heart failure and cyanosis[5]. Our patient was diagnosed very early, at the age of 2 days.

Due to high pulmonary blood flow, pulmonary hypertension is an inevitable complication of hemitruncus arteriosus[6]. If untreated, pulmonary hypertension becomes irreversible, and the lesion will be inoperable. The mortality of hemitruncus arteriosus is extremely high, with over 30% and 70% of untreated infants dying at 3 and 12 months, respectively[6]. Early correction is associated with a better prognosis[6]. Our neonate had severe pulmonary hypertension at diagnosis. She underwent surgical correction at 28 days and had been living a normal life since then. Early clinical suspicion and timely investigation and referral were key to the successful management of the case described here.

In conclusion, early clinical suspicion, accurate diagnosis and timely corrective surgery are important for life-threatening congenital heart diseases in neonates.

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