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A Case of Reverse Differential Cyanosis with the Combination of Transposition of the Great Arteries and Aortic Coarctation

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Abstract

Reverse differential cyanosis is a very rare symptom characterized by cyanosis in the upper extremities without cyanosis in the lower extremities (high SpO2). The combination of aortic coarctation and D-loop transposition of the great arteries is the most common cause of this condition. Here, we present a case of transposition of the great arteries associated with aortic coarctation, which manifested as reverse differential cyanosis immediately after birth.

Keywords: dTGA; Fetal diagnosis; Pulmonary hypertension; Ductus arteriosus

Case Presentation

40+4-week-old male infant, weighing 345 g, was born via caesarean section to a 27-year-old mother (gravida 0, para 0). The mother did not attend prenatal check-ups during the pregnancy. The Apgar scores were 9 and 9 at 1 and 5 minutes after birth, respectively. Immediately after birth, nasal breathing with grunting and retractions was observed, along with an SpO2 of 60%. Oxygen was administered via nasal cannula [1,2].

On physical examination, a 2/6 systolic murmur was heard at the mesocardial focus. Upper extremity pulses were palpable, and the blood pressure was measured at 80/45 mmHg. However, femoral, posterior tibial, and dorsalis pedis artery pulses were absent, and blood pressure could not be measured in the lower extremities. Within the first hour after birth, SpO2 was 78% in the upper extremities and 88% in the lower extremities. A chest X-ray revealed a cardiothoracic ratio of 54%. The electrocardiogram (ECG) showed a sinus rhythm with a rate of 150 bpm, right axis deviation, PR interval of 100 ms, QTc interval of 420 ms, and no ST-T changes. Transthoracic echocardiography revealed that the pulmonary veins drained into the left atrium and through the mitral valve into the left ventricle; however, the pulmonary artery arose from the left ventricle. The aorta and pulmonary artery ran parallel to each other (D-loop transposition of the great arteries). Additionally, a coarctation of the aorta measuring 2-3 mm was found just below the subclavian artery. The ductus arteriosus was patent, with a shunt from the pulmonary artery to the aorta. A secundum Atrial Septal Defect (ASD) measuring 4 mm with left-to-right shunting was also present. The patient was diagnosed with D-loop transposition of the great arteries and aortic coarctation.

Before surgery, the patient's respiratory rate was 52 breaths per minute, and SpO2 was 84% in the right upper extremity and 94% in the lower extremities, reflecting reverse differential cyanosis. A repeat echocardiogram showed no ventricular septal defect on the parasternal long-axis view. The aorta and pulmonary artery ran parallel and originated from the right and left ventricles, respectively (Figures A,B). Coarctation of the aorta at the level of the interruption was present, and blood flow from the pulmonary artery to the aorta was observed (Figures C,D).

To maintain the patency of the ductus arteriosus, a prostaglandin infusion was started, and the patient was transferred to a center with paediatric cardiovascular surgery for necessary interventions. The patient underwent balloon atrial septostomy, but unfortunately passed away due to acute renal failure and Disseminated Intravascular Coagulation (DIC) while awaiting surgery.

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Discussion

Differential cyanosis involves a pink appearance in the right upper extremity while cyanosis is prominent in both lower extremities [3]. This condition is observed in pulmonary artery hypertension along with a Patent Ductus Arteriosus (PDA) [3]. Deoxygenated blood in the pulmonary artery passes through the patent ductus arteriosus, empties into the aorta, and flows to the lower half of the body. The upper half of the body continues to receive oxygenated blood from the left ventricle. If the patent ductus arteriosus enters below the left subclavian artery, the left arm will appear pink; if it enters above the left subclavian artery, the left arm will appear cyanotic. In neonates, differential cyanosis indicates persistent pulmonary hypertension and left heart abnormalities [3,4].

In reverse differential cyanosis, the arms are more cyanotic than the legs. This occurs because the right ventricle directs lower oxygen-saturated blood to the upper extremities. Cyanosis is seen in the upper extremities due to the low oxygenation of blood in the aorta, which is caused by right-to-left shunting at the atrial level in cases of Pulmonary Hypertension (PH) or by the direct origin of the aorta from the right ventricle in cases of transposition of the great arteries (TGA). In the presence of aortic coarctation, the passage of blood with a higher oxygen concentration occurs from the pulmonary artery into the descending aorta via the PDA, distal to the left subclavian artery. Therefore, the lower extremities have higher oxygen saturation than the upper extremities, and cyanosis is not observed [4].

In normal fetal circulation, the pulmonary artery carries blood with 50% oxygen saturation [5]. In infants with transposition of the great arteries (TGA), the pulmonary artery carries blood with a higher oxygen saturation (around 72%) [6]. As a result, the pulmonary artery in TGA dilates and carries three times more blood compared to a normal fetus. TGA manifests as prolonged pulmonary hypertension after birth. Due to the large amount of blood flowing into the pulmonary artery during the fetal period, pulmonary hypertension can be seen as left ventricular pressure exceeding right ventricular pressure; however, pulmonary hypertension was not observed in our case.

In our case, due to D-loop transposition of the great arteries (TGA), the aorta was delivering oxygen-poor blood from the systemic venous circulation to the systemic circulation, leading to cyanosis in the upper extremities. Additionally, there was severe coarctation immediately distal to the left subclavian artery. Blood with high oxygen levels was passing through the PDA from the pulmonary artery into the descending aorta, distal to the coarctation segment. Therefore, cyanosis was not present in the lower extremities.

Conclusion

Although differential cyanosis is quite rare in newborns, it usually occurs in clinical conditions that can lead to high mortality and morbidity. Identifying these conditions during the fetal period, when possible, and performing the necessary interventions early and quickly can prevent potential complications.

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