



A Case of Dextrotransposition of the Great Arteries Type I with Reversed Differential Cyanosis

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Abstract

Transposition of the great arteries type I is a severe congenital heart disease that induces serious cyanosis immediately after birth and death within 24 hours, unless proper treatment is administered. A few cases have presented with reversed differential cyanosis, manifesting as separated cyanosis with high SpO₂ values in the lower limbs. However, there have been few reports of survivors of transposition of the great arteries type I presenting with reversed differential cyanosis. We experienced a case of transposition of the great arteries type I presenting with reversed differential cyanosis immediately after birth. The infant was urgently transported because of postnatal SpO₂ of 40% in the upper limbs and 90% in the lower limbs. The echocardiographic diagnosis was transposition of the great arteries type I with a narrow foramen ovale. We immediately performed balloon atrioseptostomy, and the reversed differential cyanosis was improved. The infant seems to have presented with reversed differential cyanosis because of the foramen ovale narrowing and complicating pulmonary hypertension due to fetal circulation and characteristic of transposition of the great arteries type I. Congenital heart disease, presenting with reversed differential cyanosis, is a clinical condition requiring emergency management.

Keywords

- ▶ dTGA
- ▶ foramen ovale
- ▶ fetal diagnosis
- ▶ pulmonary hypertension
- ▶ ductus arteriosus

Reversed differential cyanosis is characterized by separated cyanosis with high SpO₂ values in the lower limbs and is a rare symptom.¹ It is observed in transposition of the great arteries type I complicated with aortic coarctation and total anomalous pulmonary venous return.² We, herein, report a severe case of transposition of the great arteries type I without aortic coarctation that presented with reversed differential cyanosis immediately after birth; the reversed differential cyanosis was improved by emergency balloon atrioseptostomy.

Case

A female infant was born at 40 weeks' gestation to a 28-year-old woman, gravida 0, para 0, who had never had children. No

abnormality was observed during pregnancy. The patient weighed 2,830 g (−0.4 standard deviations), and the Apgar's score was 7 and 8 at 1 and 5 minutes, respectively, after birth. Oxygen administration was started because she presented with severe cyanosis of SpO₂ at 40% and nasal breathing immediately after birth. She was diagnosed with transposition of the great arteries type I by echocardiography and transferred to our hospital. Regarding the patient information, the patient had transposition of the great arteries type I and presented with reversed differential cyanosis (the SpO₂ at 2 hours and 40 minutes after birth was 60% in the upper limbs and 90% in the lower limbs). We suspected transposition of the great arteries type I based on the narrowing of the foramen ovale and hospitalized to prepare her for emergent balloon atrioseptostomy. At hospitalization, the respiratory rate was 37/min, and the SpO₂ reflected reversed differential cyanosis with a value of 60% in the upper right limb and 94%

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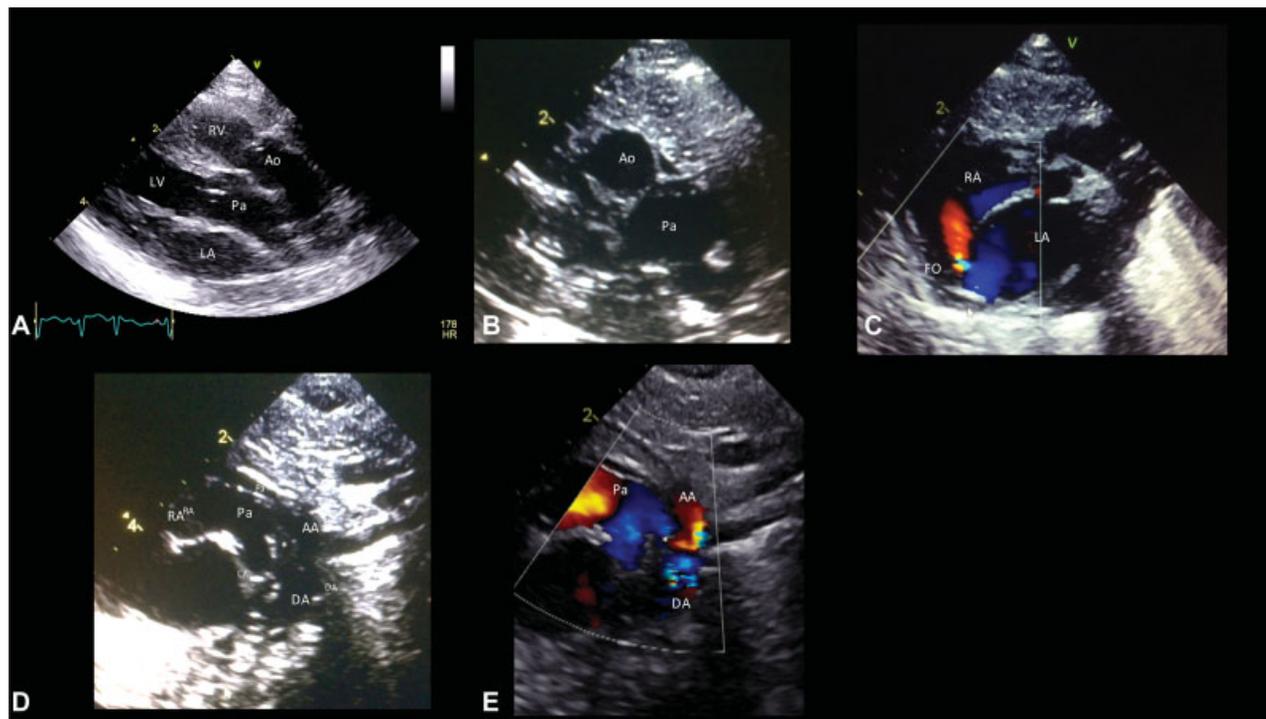


Fig. 1 (A, B) Given that the aorta and pulmonary artery were parallel in the long-axis image and originate from the right and left ventricle, respectively, we concluded that this was a case of transposition of the great arteries type I. (C) Interatrial traffic could not be identified by the B mode method, and slow blood flow was observed between the atria by the color Doppler method, so we suspected that the foramen ovale was close to closure. (D) Although there was no constriction in the aorta, the arterial duct was narrow, and blood flow from the pulmonary artery into the aorta was observed. (E) The narrow aortic section revealed blood flow traveling backward into the ascending aorta.

in the lower limbs. Chest X-ray showed a cardio thoracic ratio of 54%, and no abnormal shadow was observed in the lung field. Echocardiography, immediately after hospitalization, showed no ventricular septal defect on a parasternal long axis view, and the aorta and pulmonary artery were parallel and originated from the right and left ventricles, respectively (►Fig. 1A and B). The foramen ovale could not be identified by the B mode method, and slow blood flow was observed between the atria by the color Doppler method, so we suspected closure (►Fig. 1C). Although there was no aortic coarctation, the arterial duct was narrow, and blood flow from the pulmonary artery to the aorta was observed (►Fig. 1C and D). A reversed blood flow to the ascending aorta was observed from the aortic narrow section (►Fig. 1E). We promptly performed balloon atrioseptostomy, and the difference in the SpO₂ between the upper and lower limbs was improved. The right ventricular pressure after balloon atrioseptostomy was 45 mm Hg, while that in the left ventricle was 50 mm Hg, and the mean pulmonary venous wedge pressure was 27 mm Hg. Pulmonary hypertension was observed (►Table 1).

Discussion

Differential cyanosis is a clinical manifestation of pink upper limbs and cyanotic lower limbs and appears in congenital heart disease with pulmonary hypertension and arterial duct. When aortic coarctation is further complicated, blood

with high-oxygen saturation flows from the left ventricle to the upper limbs, and low-oxygen saturated blood flows backward from the right ventricle through the arterial duct and into the lower limbs. In cases of reversed differential cyanosis, the situation is inverted, where patients are presenting with pink lower limbs and cyanosis of the upper limbs. Reversed differential cyanosis is rare but can appear in

Table 1 Results of cardiac catheterization after balloon atrioseptostomy

Site	Pressure systole/diastole (mean) mm Hg	Oxygen saturation (%)
Superior vena cava	4/2 (2)	69.5
Right atrium	4/3 (2)	81.2
Inferior vena cava	3/1 (2)	75.3
Right ventricle	45/5	99.2
Left ventricle	50/5	99.2
Left atrium	4/2 (8)	99.7
Right upper pulmonary vein	9/6 (8)	–
Right upper pulmonary vein wedge	34/25 (27)	–
Aorta	43/24 (32)	95.6

cases of transposition of the great arteries type I with aortic coarctation and total anomalous pulmonary venous return.

Because the right ventricle diverts blood with low-oxygen saturation to the upper limbs and the left ventricle diverts blood with high-oxygen saturation to the lower limbs, reversed differential cyanosis occurs in transposition of the great arteries type I with coarctation of the aorta. Although the present case had transposition of the great arteries type I without coarctation of the aorta, the patient presented with significant reversed differential cyanosis. In normal fetal circulation, the pulmonary artery carries blood with 50% oxygen saturation.³ In the circulation of infants with transposition of the great arteries type I, the pulmonary artery carries blood with a high oxygen saturation (around 72%).⁴ As a result, the pulmonary artery of fetal transposition of the great arteries type I cases is dilated and carries thrice as much blood as a normal fetus.⁴ Transposition of the great arteries type I presents with prolonged pulmonary hypertension after birth, as a large amount of blood flows to the pulmonary artery during the fetal period. In the present case, as well, the left ventricular pressure showed pulmonary hypertension, exceeding the right ventricular pressure, although these data were obtained via a catheter after balloon atrioseptostomy. For this reason, the blood in the arterial duct flowed backward from the pulmonary artery to the descending aorta on echocardiography after birth.

The present case had transposition of the great arteries type I with narrowing of the foramen ovale. According to the American Heart Association statement, transposition of the great arteries type I is classified as a severe fetal heart disease with a prognosis that can be expected to be improved by prompt treatment after birth.⁵ According to the information available when hospitalization was requested, in addition to having transposition of the great arteries type I, the patient also had reversed differential cyanosis (which was treated with balloon atrioseptostomy), and closing of the foramen ovale was also suspected because the right upper limb SpO₂ was 60% which was very low. In the absence of mixing of arterial blood from the left tuft at the foramen oval hole, the venous blood flows directly into the aorta, thereby reducing the SpO₂ of the upper limb. The fetal pulmonary blood flow in cases of transposition of the great arteries type I is thrice that in a normal fetus, so the left atrial pressure rises, making the foramen ovale more likely to close than in a normal fetus. Therefore, observing the foramen ovale in the fetal stage is important.

Transposition of the great arteries type I, in the present case, could not be diagnosed at the fetal stage; we should, therefore, make note that cases of transposition of the great arteries type I with a low SpO₂ value in the upper limb may have a foramen ovale that is close to closure.

Yamamoto et al reported that a brain-sparing effect was observed in transposition of the great arteries type I, reflecting the decreased oxygen saturation of the cerebral blood flow during the fetal period.⁶ The brain-sparing effect occurs when the blood flow reverses in the aorta as a defensive

reaction to reduce the blood flow to the limbs and abdominal organs and increase the blood flow to the brain. In the present case, the reversal of the blood flow from the aortic stenosis to the ascending aorta was observed with echocardiography after birth (–Fig. 1E). However, our search revealed no similar reports of blood flowing backward to the ascending aorta in postnatal echocardiography. We suspect that the brain-sparing effect in the present case continued after the occurrence of reversed differential cyanosis continued, even after the occurrence of high pulmonary vascular resistance and decreased oxygen saturation of cerebral blood flow.

Congenital heart disease with reversed differential cyanosis is limited to total anomalous pulmonary venous return and transposition of the great arteries. Furthermore, in transposition of the great arteries, the upper extremity SpO₂ reflects the severity. Therefore, if reversed differential cyanosis is suspected, we should pay attention to the SpO₂ value in the upper limbs and respond urgently when it is low.

In addition, the blood flowing backward into the aorta reflects a decrease in the oxygen saturation of the cerebral blood flow, and an urgent response is necessary to ensure a good cranial neurological prognosis.

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Conflicts of Interest

None.

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