Reversed Differential Cyanosis

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Deeper cyanosis of the upper than of the lower part of the body, or reversed differential cyanosis, is a rare but important physical sign. It has been observed in patients with complete transposition of the great vessels with or without hypoplasia, coarctation, or interruption of the aortic arch (Hamburger, 1937; Hecko, 1948; Buckley *et al.*, 1965). This case report concerns a child who had transposition of the great vessels associated with interruption of the arch of the aorta distal to the origin of the left subclavian artery and who showed reversed differential cyanosis.

Case Report

S.W., a 23-month-old white boy, was first admitted to hospital at the age of 3 weeks with a history of cyanosis since birth, failure to thrive, tachycardia, and tachypnoea. Pregnancy and delivery had been normal and the birthweight was 3402 g. (7 lb. 8 oz.). On examination he was cyanosed and showed moderate respiratory distress. The cyanosis was symmetrical but deeper over the face and both arms than over the lower part of the body. The heart rate was 170 and the respiratory rate 40 a minute. The femoral pulses were easily palpable. The blood pressure was 95/40 mm. Hg in the legs and 85/35 mm. Hg in the right arm. The heart was not enlarged clinically and there were no thrills. On auscultation there was a harsh grade 3/4 ejection systolic

murmur, loudest in the third and fourth left intercostal spaces parasternally. There was no diastolic murmur. The second heart sound was of normal intensity and physiologically split. The liver was palpable 3 cm. below the right costal margin and was not pulsatile. Haemoglobin was 17 g./100 ml.

The electrocardiogram showed left axis deviation, and right atrial and right ventricular hypertrophy. Normal heart size with a narrow pedicle and increased pulmonary vasculature were seen on the x-ray film.

Cardiac catheterization was performed initially at the age of 3 weeks and was repeated at 23 months: the findings were similar on each occasion. Neither great artery was entered. There was a left-to-right shunt at ventricular level with right ventricular hypertension (Table). Oxygen saturation in the femoral artery was 52 per cent and in the brachial artery 32 per cent, confirming the presence of reversed differential cyanosis. Selective angiocardiology was performed on both occasions, the contrast medium being injected into the right ventricle at the first investigation and into the left atrium at the second. These showed complete transposition of the great vessels, interruption of the aortic arch distal to the origin of the left subclavian artery, and right-to-left shunt at ventricular and pulmonary artery levels. The descending aorta was filled from the pulmonary artery through a patent ductus arteriosus (Fig. 1 and 2).

Cardiac failure was treated with digoxin and diuretics. This therapy has been continued and, though he responded well initially, frequent respiratory tract infections have persisted. He is now over 2 years old, but has not thrived and his development is retarded.

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TABLE
CARDIAC CATHETERIZATION AT 23 MONTHS*

	Position of catheter					
	Superior vena cava	Right atrium	Right ventricle	Left atrium	Femoral artery	Brachial artery
Oxygen saturation (%)	30	30	44	99	52	32
Pressure (mm. Hg)	—	6/2	72/2	9/3	—	—

* Critical condition of the infant precluded a more detailed study.

FIG. 1.—Selective right ventricular angiogram. Antero-posterior (a) and lateral (b) views. RV, right ventricle; A, ascending aorta; PA, pulmonary artery; DA, descending aorta; D, ductus. In lateral view, note anteriorly placed aorta being filled from right ventricle, arrow pointing the site of interruption of the arch of the aorta, and the descending aorta being filled from the pulmonary artery through the ductus arteriosus.

FIG. 2.—Selective left atrial angiogram. Antero-posterior (a) and lateral views (b). LA, left atrium; LV, left ventricle. In lateral view, note prominent pulmonary artery being filled from left ventricle and the dye passing to the descending aorta through the patent ductus arteriosus.

Discussion

Reversed differential cyanosis may occur with complete transposition of the great vessels associated with pulmonary hypertension and a patent ductus arteriosus. Oxygenated blood from the left ventricle passes into the pulmonary artery and through the patent ductus arteriosus to the descending aorta, while systemic venous blood flows from the right ventricle into the ascending aorta and its branches (Buckley *et al.*, 1965). Hypoplasia, coarctation, or atresia of the aortic arch greatly increases the degree of reversed differential cyanosis (Moller and Edwards, 1965). With complete interruption of the aortic arch distal to the origin of the left subclavian artery, cyanosis in both arms is intense and symmetrical as in our case (Buckley *et al.*, 1965). Poor intracardiac mixing of blood due to small defects or equal pulmonary and systemic resistance may be expected further to intensify the degree of reversed differential cyanosis.

It must be noted, however, that in most cases the associated intracardiac defects allow a high degree of mixing of the blood in the heart, and in the absence of obstruction to blood flow from the ascending to the descending aorta sufficient desaturated blood passes to the descending aorta to make the difference in cyanosis between the upper and the lower extremities slight and difficult or impossible to detect clinically (Bowers, Schiebler, and Krovetz, 1965). Proximity of the left subclavian artery to the ductus may allow retrograde flow of oxygen-

ated blood into this artery, so that cyanosis is less intense in the left than in the right arm.

Angiocardiography provides the best means of establishing the diagnosis. The complex nature of the anomaly in this child precludes surgical correction at present and the prognosis is poor. This child was over 2 years when last seen, but his physical development was grossly retarded.

Summary

A case of transposition of the great vessels with coarctation of the aorta and a patent ductus arteriosus is described, and reversed differential cyanosis is discussed.

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