COARCTATION OF THE AORTA IN ONE OF IDENTICAL TWINS

C. D. DRIVER, M.B., CH.B. (CAPE TOWN), and PETER V. SUCKLING, M.D. (LOND.), M.R.C.P., D.C.H.
Victoria Hospital, Wynberg, Cape

Since identical twins have the same genetic structure, anatomical differences between one identical twin and the other must have an environmental cause, from factors operating either *in utero* or in postnatal life. In the present case, it must be presumed that the lesion occurred during intra-uterine life because of the congenital nature of the lesion and age of the child.

This appears to be the first case to be reported of coarctation of the aorta in one of monozygous twins.

CASE HISTORY

Cheryl is a Coloured* female infant born on 13 October 1958, and was brought to the out-patient department of this hospital on 21 December 1958 because of a mild gastro-enteritis. A cardiac murmur was discovered, and on further examination the signs of coarctation of the aorta. Since she proved to be one of twins, her sister was sent for and the two admitted for investigation.

The twins come 7th in the family, the 6 older sibs all being singletons and in good health. There is no history of twins on the mother's side of the family, but the father has two cousins both of whom have twins, one pair being reported as identical.

The pregnancy was normal, without overt maternal infection, threatened miscarriage or antepartum haemorrhage.

Delivery. The twins were delivered at home, and the midwife noted that there was only one placenta, which was large. Cynthia was born first, by the vertex, and weighed 5 lb. (2·3 kg.). She was followed 20 minutes later by the patient Cheryl, born by the breech and weighing 4¾ lb. (2·2 kg.).

Postnatal period. The twins were breast-fed for the first 4 months, but were then put on to a half-cream milk mixture owing to the failure of the mother's milk supply. Both sucked well, but Cheryl has remained slightly smaller than her sister.

Physical Examination at 5½ months

At the age of 5½ months Cheryl's weight was 6 lb. (2·7 kg.) and she was 23 inches (58 cm.) in length. Cynthia weighed 6 lb. 3 oz. (2·8 kg.) and her length was 23½ inches (59 cm.). They

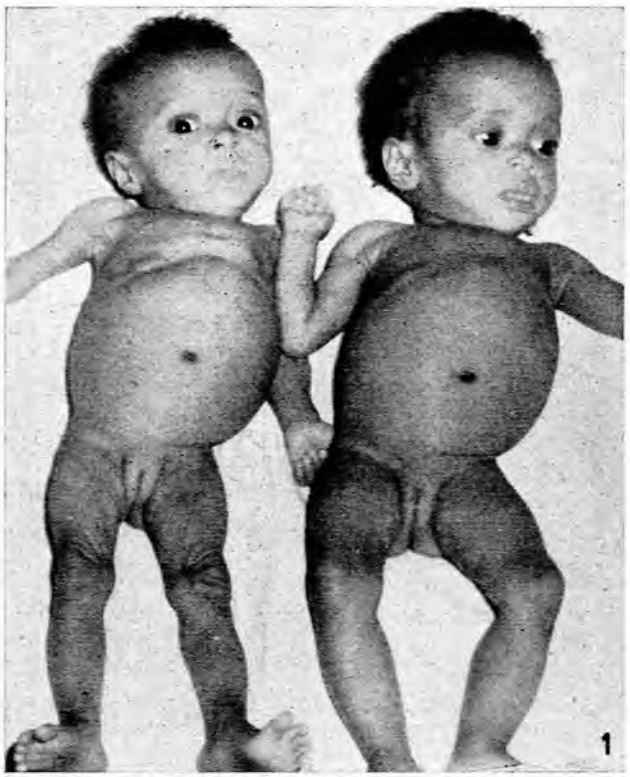

Fig. 1. Patient Cheryl (left) and her twin sister Cynthia.

* The Coloured race is derived from a mixture of European with Malay, Hottentot, Bantu or Bushman stock.

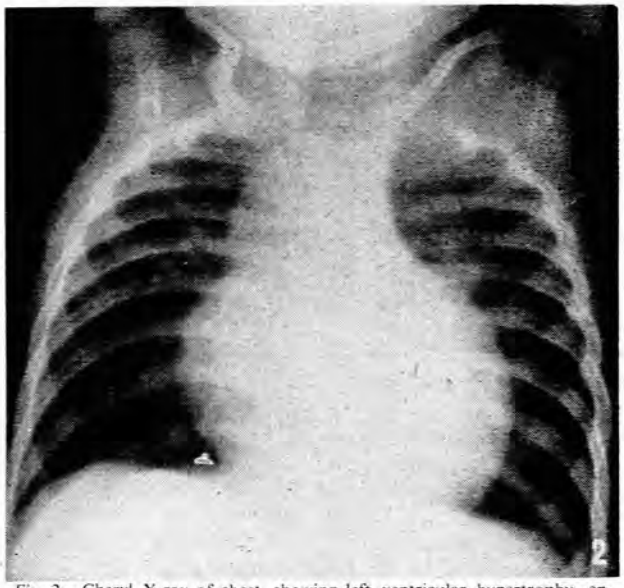

Fig. 2. Cheryl X-ray of chest, showing left ventricular hypertrophy, enlargement of ascending aortic shadow, and well-marked pulmonary bay.

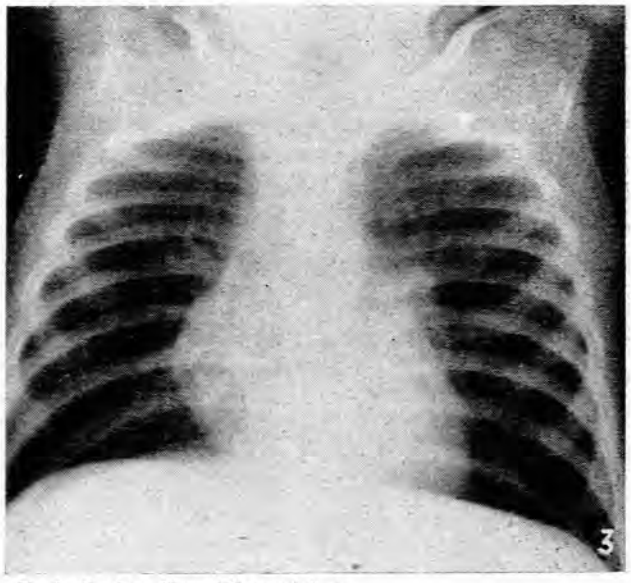

Fig. 3. Cynthia. Normal X-ray of chest.

were very similar in appearance, but it was noted that Cheryl's lower limbs appeared smaller than those of her sister (Fig. 1).

Neither was cyanotic nor exhibited clubbing of the fingers, and there was no evidence of cardiac failure. Except for the marasmus and the physical signs in Cheryl's cardiovascular system, no abnormalities were found (Table I and Figs. 2 and 3).

TABLE I. THE CARDIOVASCULAR SYSTEM

	Cheryl	*Cynthia*
Radial pulses	115/min., full regular, collapsing.	110/min., regular normal volume and tension.
Femoral pulses	Not palpable.	Palpable, and of comparable fullness and tension with the radials.
Blood pressure		
Arms	125/45 mm. Hg.	85/40 mm. Hg.
Legs	Not recordable.	85/40 mm. Hg.
Heart		
Apex beat	4th left intercostal space, mid-clavicular line, left ventricular in type.	4th left intercostal space, 1 cm. medial to the mid-clavicular line, normal in character.
Sounds	Normal at apex. P2 split.	Normal. P2 split.
Murmur	Basal holosystolic murmur, conducted into neck vessels.	None.
Collateral circulation	Not demonstrated.	—
Electrocardiogram	Left ventricular hypertrophy.	Normal.
X-ray (Figs. 2 and 3)	Cardiac hypertrophy. Enlargement of ascending aortic shadow, and well-marked pulmonary bay.	Normal.

Evidence of Monozygosity

There was only one large placenta, as vouched for by the midwife.

The features are remarkably similar, the hair is alike in colour and whorl, and the eyes are alike. The ears and lips are similar. The twins have identical blood groups, viz. Group B, ccDee, NNS, K negative, Duffy positive.

The finger prints are dissimilar; Cheryl's are of a whorled nature, whereas the basic pattern of Cynthia's is looped.

DISCUSSION

Narrowing or coarctation of the aorta may occur at any site, but in the vast majority of cases the stenosis lies immediately beyond the origin of the left subclavian artery in close relationship to the ligamentum arteriosum. Two main types are described, as follows:

1. *Infantile type.* Here the ductus remains patent and enters the aorta below the stenosis. The pressure in the distal aorta is so low that venous blood passes from the pulmonary artery into the thoracic aorta, resulting in cyanosis of the lower half of the body, whilst the upper half remains pink. Other congenital abnormalities are frequent, and the condition is regarded as being incompatible with life for more than a few weeks after birth.

2. *Adult type.* In this, by far the commoner, type the ductus is usually obliterated or, if patent, opens into the aorta above the stenosis. The present case is of this type.

It has been shown that the arch of the aorta is formed by the ventral root of the left 4th arch, by the 4th arch itself, and by the dorsal root of the left 4th arch. The first part of the descending aorta and the ductus arteriosus are formed from the left 6th arch. These developments take place between the 5th and 7th week of intra-uterine life, i.e. between the 9 mm. and 15 mm. stages of embryonic growth. It is probable, therefore, that coarctation of the aorta has its origin at this early stage of foetal development. In the present case, no aetiological factor may be demonstrated for Cheryl's coarctation.

The occurrence of congenital heart disease in one of a pair of twins has frequently been recorded in the literature, but no record of coarctation in the one twin has been found. Jones[1] records 11 cases of congenital heart disease, of which 6 were regarded as being definitely monozygotic and 4 probably so. A further 13 cases, including tetralogy of Fallot, dextrocardia with pulmonary stenosis, atrial septal defect, ventricular septal defect, pulmonary stenosis with a normal aortic root, and primary endocardial fibro-elastosis, may be added from the paper by Uchida and Rowe.[2]

SUMMARY

A case of coarctation of the aorta in an identical twin is reported. No aetiological factor could be demonstrated.

We wish to thank the Medical Superintendent for permission to publish, and Dr. L. Vogelpoel for the ECG report.

ADDENDUM

The patient Charyl died suddenly before an arrangement could be made for her admission to hospital for an operation to correct the coarctation. An autopsy was carried out by Dr. W. P. Mulligan, whose report is as follows:

An autopsy was carried out on this child on 22 January 1960 about 36 hours after death. The body is that of a thin underweight child. PM lividity moderate. No rigor mortis noted. Moderate oedema noted.

Cardiovascular system. The heart is moderately enlarged, with marked left ventricular hypertrophy (wall 1 cm. thick). Slight right ventricular hypertrophy noted. Valves normal. No bicuspid aortic valve. A coarctation of the aorta is noted in the arch about 3 cm. from the aortic ring. Here the vessel is 1·2 cm. in diameter. Proximally the aorta is 2·5 cm. in diameter. Coarctation is at the site of the closed ductus.

Dilated arteries coming from the aorta proximal to the coarctation are noted. They are the two innominates coming off together and the left subclavian. The intercostals below the coarctation are small and so are the common iliacs. The foramen ovale is patent, with an opening only 2 mm. in diameter.

Respiratory system. Left lung 70 g. Right lung 85 g. There is collapse of the whole left lower lobe, with bronchial mucus in excess. Subpleural petechial haemorrhages noted.

Gastro-intestinal system. Moderate fatty change in liver.

Genito-urinary system. Normal.

Brain. Normal. No haemorrhage or aneurysm.

Conclusion. Death appears to be due to bronchitis with collapse of the lower lobe of the left lung in a malnourished child with a coarctation of the aorta.

REFERENCES

1. Jones, H. E. (1958): Arch. Dis. Childh., 33, 342.
2. Uchida, I. A. and Rowe, R. D. (1957): Amer. J. Hum. Genet., 9, 133.