Truncal valve stenosis

Richard H. Burnell, Gerald McEnery, and Graham A. H. Miller¹

From the Paediatric and Cardiac Departments, Brompton Hospital, London S.W.3

A case is presented of truncus arteriosus with truncal valve stenosis, demonstrated on haemodynamic findings in life and confirmed subsequently at necropsy. Though analogies may be drawn with the relatively common aortic valve stenosis of infancy, this appears to be the first demonstration in life of truncal valve stenosis.

Structurally abnormal valve cusps in truncus arteriosus have been recognized for many years (Roos, 1935), but it appears that Deely, Hagstrom, and Engle (1963) were the first to associate these with truncal valve insufficiency. The opposite occurrence, that of truncal valve stenosis, does not appear to have been diagnosed previously in life and is the reason for this case report.

Case history

The patient was the first child of a 34-year-old woman and was born after 38 weeks' gestation (birthweight 2.3 kg.). Except for a systolic murmur, noted immediately after birth, no abnormalities were detected until the 8th day of life, when she had a transient cyanotic attack and was thought subsequently to be in cardiac failure. Digitalis and diuretic therapy was begun and she was transferred to Brompton Hospital.

On examination she exhibited tachypnoea and tachycardia, but no overt cyanosis. There was slight oedema of the face, hands, and feet, and obvious hepatomegaly. The respiratory system was clear. The cardiovascular system showed diminution of all peripheral pulses, a systolic thrill, and grade 5/6 rough ejection systolic murmur maximal in the upper right intercostal spaces, and an apical gallop sound. The second heart sound was single.

Investigations Haemoglobin 13.6 g./100 ml. Serum electrolytes and urea normal. Electrocardiogram was interpreted as showing conspicuous right ventricular hypertrophy. ST depression and T inversion were present from V2-V7. Chest x-ray showed moderate cardiomegaly and increased pulmonary vascularity.

Progress Despite some weight loss in response to treatment, the baby continued to be very tachypnoeic. On the 13th day of life, right and left heart catheterization was performed (Table). The venous catheter crossed from the right ven-

tricle into the aorta, but not into the pulmonary artery. The retrograde arterial catheter passed from the aorta to the left ventricle. The ventricular pressures were similar, and a 70 mm Hg aortic systolic gradient was present.

A right ventricular biplane angiogram was interpreted as showing a ventricular septal defect with right-to-left shunting into the aorta, no delineation of a pulmonary outflow tract, and equal filling of the right and left pulmonary arteries. It was not possible to be certain how the pulmonary arteries filled, but truncus arteriosus was the suggested diagnosis.

Because of the baby's poor condition, an attempt to perform valvotomy was begun on the 17th day of life. The baby succumbed shortly after induction of anaesthesia.

Necropsy was performed 36 hours after death. Significant anomalies were confined to the cardiovascular system. The heart was enlarged by right atrial dilatation and hypertrophy of both ventricles, particularly obvious in the right ventricle. There was a high ventricular septal defect (0.5 cm. in diameter) over which rose a single large truncal vessel, giving origin to the right and left coronary arteries. 1 cm. above the truncal valve there were separate orifices of the right and left pulmonary arteries. The truncal vessel then continued as ascending aorta with a normal distribution of the brachiocephalic vessels. The aortic arch was left-sided (Fig.).

The truncal valve was quadricuspid, each leaflet being mostly composed of an irregular

TABLE *Haemodynamic data*

Site	Pressures (mm Hg)	O ₂ saturation (%)
Superior vena cava	—	34
Inferior vena cava	—	36
Right atrium	Mean = 8	53
Low right ventricle	130/4-16	69
High right ventricle	130/4-16	81
Left atrium	Mean = 12	98
Left ventricle	140/14	98
Aorta	73/44	84
Axillary artery	73/44	84

¹ Correspondence should be addressed to Dr. G. A. H. Miller, Cardiac Department, Brompton Hospital, London S.W.3.

FIG. Truncal valve region. Origin of left pulmonary artery not visible. AA, ascending aorta; MV, anterior leaflet of mitral valve; RPA, origin of right pulmonary artery; TV, truncal valve; VSD, ventricular septal defect.

firm nodule of pinkish myxomatous material. There was fusion of all 4 commissures, leaving a small valve orifice. Before the valve was opened the orifice measured no more than 2.5 mm. in diameter. There was fibrous continuity of mitral and truncal valves. A patent foramen ovale was present and was considered to be valve-incompetent.

The pathological findings were Type A₂ truncus arteriosus of Van Praagh and Van Praagh (1965) with truncal valve stenosis.

Discussion

Thickened nodular valve cusps in congenital aortic valve stenosis of infancy have been well described (Hastreiter *et al.*, 1963) and macroscopically at least resemble those seen in some cases of truncus arteriosus. This is not surprising; as Van Praagh and Van Praagh (1965) pointed out, the truncal valve embryologically is an aortic valve. However, truncal valve stenosis has not apparently been described as a clinical entity, nor yet as a pathological one, with the exception of Case 44 of Van Praagh and Van Praagh (1965). This case is labelled 'truncal valve stenosis probably', without further comment.

It seems reasonable to suppose that the lesion might usually be lethal *in utero*, but since survival past term is possible it is felt

that some features of this case are worth emphasizing.

Firstly, the presence of strikingly diminished peripheral pulses and a harsh ejection systolic murmur maximal in the upper right intercostal spaces led several observers to diagnose aortic valve stenosis, despite the electrocardiogram which did not show left ventricular hypertrophy – the rule in infantile aortic stenosis (Hastreiter *et al.*, 1963). Diminished peripheral pulses are of course exceptional in truncus arteriosus, but an aortic ejection murmur is not; a similar murmur has been described by Tandon, Hauck, and Nadas (1963) in 8 of 19 cases of truncus arteriosus without haemodynamic evidence of truncal valve stenosis. It may be that such a murmur is heard in those cases of truncus arteriosus with abnormal truncal leaflets.

Secondly, the haemodynamic findings shown in this case were those which might have been predicted for the anatomy, i.e. evidence of a mixing situation at ventricular level and both ventricles at equal pressure well above systemic pressure. A right ventricular angiogram was correctly interpreted by one radiologist as that of truncus arteriosus, though his confidence was shaken on being told subsequently of the 70 mm Hg systolic gradient. Had we paid more attention to Holmes' dictum concerning the impossible and the improbable (Doyle, 1901) a firm rather than a tentative diagnosis might have been made.

Finally, in view of the grossly abnormal truncal valve, it seems improbable that any standard surgical approach would significantly alter the outcome of this or similar cases.

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