Case reports

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Infradiaphragmatic total anomalous pulmonary venous return *Diagnosis and surgical repair in a 10-year-old child*¹

Desmond F. Duff, Michael R. Nihill, Thomas A. Vargo, and Denton A. Cooley

From The Section of Cardiology, Department of Paediatrics, Baylor College of Medicine, and Texas Children's Hospital; and The Surgical Section, Texas Heart Institute, Houston, Texas, U.S.A.

A unique case of infradiaphragmatic total anomalous pulmonary venous return in a 10-year-old girl is described. A persistent ductus arteriosus, ventricular septal defect, and large atrial septal defect were also present. Corrective surgery was performed at 10 years of age, with an excellent clinical result. The factors which permitted this unprecedented period of survival included non-obstructed drainage of the pulmonary venous blood into the inferior vena cava and a large atrial septal defect.

Infradiaphragmatic total anomalous pulmonary venous return comprises only 0.1 to 0.2 per cent of all congenital heart lesions. Usually the anomaly produces pulmonary venous obstruction, and except for the few who have had successful surgical treatment, the outcome is uniformly fatal in the first weeks or months of life (Keith, Rowe, and Vlad, 1967). This report concerns a unique patient in whom the anomaly was first diagnosed at the age of 10 years, and who then underwent successful surgery.

Case report

The patient was first admitted to Texas Children's Hospital at the age of 10 years for further evaluation of cyanotic congenital heart disease. Cyanosis, respiratory difficulty, and a heart murmur had been noted at birth, and she required intensive care, including supportive ventilation. She also had the features of Goldenhar's syndrome (Feingold, 1973). Cyanosis, exertional fatigue, dyspnoea, and growth failure persisted during infancy and childhood. Cardiac catheterizations were performed at 3 months and 5 years of age at another hospital, and it was concluded that she had an atrial septal defect, ventricular septal defect, persistent ductus arteriosus, and irreversible pulmonary vascular obstruction.

There was a large right-to-left shunt at atrial level, a bidirectional shunt at the ventricular septal defect, and a small left-to-right shunt at the persistent ductus arteriosus. The systemic arterial oxygen saturation was 70 per cent.

During mandibulo-facial plastic surgery at the age of 8 years, she had a cardiac arrest from which she was successfully resuscitated.

Examination on admission to Texas Children's Hospital revealed a thin child whose height and weight were below the third percentile for her age. She had the auriculo-facial and ocular manifestations of Goldenhar's syndrome. She was slightly cyanosed and this increased with effort. There was no clubbing of the digits. The heart rate was 104 per minute and the peripheral pulses were normal. The respiratory rate was 30 per minute. Blood pressure was 120/80 mmHg (16.0/10.6 kPa) in both arms. The jugular venous pulse was not raised. The transverse diameter of the chest was narrow, with a prominent left hemithorax and a hyperdynamic precordium. The apex beat was in the sixth left intercostal space at the anterior axillary line. A prominent right ventricular impulse was noted. There was no thrill. The first heart sound was normal; the second heart sound was split, with an accentuated pulmonary component. There was a grade 3/6 plateau systolic murmur along the left sternal border, most intense at the third left intercostal space, with wide radiation over the precordium. A short, low-pitched mid-diastolic murmur was heard at the tricuspid area. The chest x-ray (Fig. 1) demonstrated moderate cardiomegaly, a prominent main pulmonary artery segment, small aortic knob, and an

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FIG. 1 Chest x-ray at 10 years of age. The 2 m standard posteroanterior view shows moderate cardiomegaly, a prominent main pulmonary artery segment, small aortic knob, and an increase in size of the proximal and distal pulmonary vessels. In addition, there is a prominent inferior vena caval shadow.

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The electrocardiogram showed a frontal plane P wave mean axis of -30 degrees, a QRS mean axis in the

frontal plane of $+120$ degrees, an rsR' pattern in the right chest leads, and right ventricular hypertrophy with 'strain'.

The loud murmur characteristic of ventricular septal defect, with cardiomegaly and dilated distal pulmonary vessels, suggested that the patient did not have pulmonary vascular obstructive disease. The mid-diastolic tricuspid murmur suggested increased flow across the tricuspid valve. Cardiac catheterization was, therefore, repeated.

Cardiac catheterization

The results of the study are summarized in the Table. The low right atrial, right ventricular, pulmonary arterial, and systemic arterial oxygen saturations were similar and slightly lower than the left atrial sample. It was not possible to enter the pulmonary veins from either the left or right atrium. Peak systolic pressure at systemic level was recorded in the right ventricle and main pulmonary artery, with no significant change in the mean pulmonary artery pressure after the administration of oxygen, or oxygen with tolazoline. However, there was an apparent increase in the left-to-right shunt with oxygen and an associated fall in the pulmonary to systemic resistance ratio.

Cineangiography from pulmonary artery injections showed rapid flow of contrast medium through the pulmonary vascular bed with return through four pulmonary veins to a common chamber which was dorsal to the right and left atria (Fig. 2a and 2b). From here the common pulmonary venous channel descended to the epigastric region below the diaphragm but the further course was not defined. A left ventricular injection in the lateral view demonstrated a ventricle of normal size and an infracristal ventricular septal defect. Aortic root injection showed a large persistent ductus

TABLE Haemodynamic data

	O ₂ saturation (%)		Pressure (mmHg) (kPa)		100% O ₂
	Room air	100% O ₂	Basal		
Superior vena cava	57	74			
Right atrium, high	79				
Right atrium, low	85	96	$\bar{7}-\bar{8}$	$\bar{0.9}-\bar{1.1}$	
Inferior vena cava	80				
Left atrium	88	94	$\bar{7}-\bar{8}$	$\bar{0.9}-\bar{1.1}$	
Right ventricle	84		110/0, 11	14.6/0, 1.5	
Main pulmonary artery	85	95	110/60	$\bar{80}$ 14.6/8.0, 10.6	105/55 $\bar{70}$ 14.0/7.3 $\bar{9.3}$
Pulmonary artery wedge			$\bar{15}-\bar{19}$	$\bar{2.0}-\bar{2.5}$	
Left ventricle	85		110/0, 11	14.6/0, 1.5	
Femoral artery	85	95	125/70	$\bar{97}$ 16.6/9.3 $\bar{12.9}$	115/70 $\bar{89}$ 15.3/9.3 $\bar{11.8}$

$\dot{Q}_p/\dot{Q}_s$ Room air (assuming pulmonary venous O₂ sat. 95 per cent) 2.8:1
 Rp/Rs Room air 0.24:1

$\dot{Q}_p/\dot{Q}_s$ = Pulmonary to systemic flow ratio; Rp/Rs = Pulmonary to systemic resistance ratio.
 Figures in italics are pressures in kPa. Figures with bars are mean pressures.

FIG. 2 a) Angiogram: This shows the right and left pulmonary veins, and the common pulmonary venous chamber coursing below the diaphragm. Injection of contrast media was made in the right pulmonary artery. b) Artist's impression of the complete anomaly (apart from the aberrant origin of the right subclavian artery), with atrial septal defect, ventricular septal defect, persistent ductus arteriosus, and relevant oxygen saturations included. The arrows indicate the large right-to-left shunt at atrial level detected by cardiogreen dye and small left-to-right shunts demonstrated angiographically at the persistent ductus arteriosus and ventricular septal defect.

arteriosus and a left aortic arch with anomalous origin of the right subclavian artery from the descending aorta.

Surgery

Surgical correction was recommended and was performed using total cardiopulmonary bypass. The four pulmonary veins were found to enter a common venous channel which descended through the diaphragm and entered the inferior vena cava. A large anastomosis was made between the common pulmonary venous chamber and the posterior wall of the left atrium. The distal end of the pulmonary venous trunk was ligated at the level of the diaphragm. The persistent ductus arteriosus was doubly ligated. Closure of a large secundum atrial septal defect required a 'dacron' patch, as did closure of the ventricular septal defect. The latter was closed from the right atrium through the tricuspid valve. The peak systolic pressure in the right ventricle at the end of the procedure was 50 mmHg (6.7 kPa), while the systemic arterial peak systolic pressure was 95 mmHg (12.6 kPa). The postoperative course was uneventful, and at the time of discharge, she had no murmur, a smaller heart size radiographically, and improved exercise tolerance.

Discussion

In infradiaphragmatic total anomalous pulmonary venous return, the pulmonary veins coalesce to form a common chamber which is usually situated directly behind the left atrium. From this common chamber a single common pulmonary venous channel descends through the diaphragm, most commonly through the oesophageal hiatus and drains into either the portal vein, ductus venosus, left hepatic vein, or the inferior vena cava, as in this case. Between 50 and 60 per cent of patients with infradiaphragmatic total anomalous pulmonary venous return have no associated cardiac malformations except the essential interatrial communication and frequently a persistent ductus arteriosus (Burroughs and Edwards, 1960).

The uniformly poor prognosis in this anomaly is related to early severe pulmonary oedema and hypoxaemia secondary to pulmonary venous obstruction (Keith *et al.*, 1967).

Among patients with all forms of total anomalous pulmonary venous return, drainage occurs to the

inferior vena cava in only about 2 per cent, and in these patients obstruction may or may not occur. Twelve cases of pulmonary venous return to the inferior vena cava have been published, with an invariably fatal outcome in surgically untreated patients (Gott *et al.*, 1956; Behrendt *et al.*, 1972; Bharati and Lev, 1973; Bonham-Carter, Capriles, and Noe, 1969; Burroughs and Edwards, 1960; Keith *et al.*, 1967; Hastreiter *et al.*, 1962). Successful surgical repair in early infancy has been reported in 2 patients (Sloan *et al.*, 1962; Cooley and Balas, 1962). No previous patient has been reported to have lived beyond the age of 6 months without surgical intervention. It appears that our patient survived because there was no obstruction to pulmonary venous return at the site of anomalous connexion or at the atrial level. The difference between the pulmonary artery wedge and right atrial mean pressures was presumably caused by the increased pulmonary blood flow rather than by obstruction to pulmonary venous return (Kuramoto and Rodbard, 1962).

Raised pulmonary artery pressure is invariably present in the obstructive form of total anomalous pulmonary venous return and is usually of a severe degree, being equal to or greater than systemic pressure (Gathman and Nadas, 1970). In patients with the non-obstructive form, pulmonary artery pressure is not always raised, being related to the pulmonary flow and the response of the pulmonary vascular bed.

Though severe proliferative intimal changes in the small pulmonary arteries have been described in infants with non-obstructive total anomalous pulmonary venous return they are rare (Levy *et al.*, 1965; Jensen and Blount, 1971). The usual pathology is medial hypertrophy, grade I of Heath and Edwards classification (Heath and Edwards, 1958). The severity of the pulmonary vascular lesion increases with age, and intimal lesions in arterioles may be significant in older children and adults (Lucas and Schmidt, 1968). The haemodynamic data in our patient (Table) suggest that pulmonary vascular disease may not be too advanced because of (1) the large left-to-right shunt, both on room air and oxygen; (2) pulmonary resistance less than 30 per cent of systemic; (3) the rapid flow through the pulmonary circulation at angiography; and (4) peak systolic right ventricular pressure falling to 50 per cent of systemic pressure at the end of operation. The right-to-left shunt at atrial level was caused by obligatory total mixing of systemic and pulmonary venous blood in the right atrium rather than by raised pulmonary vascular resistance.

Cardiac anomalies are described in 20 to 50 per cent of cases of Goldenhar's syndrome (Feingold,

1973; Greenwood *et al.*, 1974). This case represents the first report of a patient with Goldenhar's syndrome in whom total anomalous pulmonary venous return was present.

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Requests for reprints to Dr. Desmond F. Duff, Department of Pediatric Cardiology, Texas Children's Hospital, 6621 Fannin, Houston, Texas 77025, U.S.A.