Infradiaphragmatic total anomalous pulmonary venous return

Review of clinical and pathological findings and results of operation in 28 cases¹

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Twenty-eight cases of infradiaphragmatic total anomalous pulmonary venous return are presented, 17 without associated complex intracardiac anomalies (group A), and 11 with additional complex lesions (group B). The anomalous site of connection was to the portal vein in 19 cases (68%), to the inferior vena cava in 4 (14%), the ductus venosus in 2 (7%), to the left hepatic vein in 2 (7%), and unknown in one. A patent foramen ovale was present in 82 per cent of cases in group A and 40 per cent in group B and was frequently associated with a small left atrium and left ventricle. Nine cases (8 in group A; 1 in group B) had surgical correction, with 3 long-term survivors. The surgical mortality was 66 per cent. The postoperative haemodynamic status of the 3 surviving patients is very satisfactory, though 1 has a residual atrial septal defect. Factors which adversely affected the surgical outcome were: (1) a critically ill infant, (2) small left atrium and left ventricle, (3) a patent foramen ovale rather than atrial septal defect, (4) systemic arterial oxygen saturation less than 70 per cent, and (5) pulmonary arterial pressure in excess of systemic arterial pressure. The mortality for the entire series was 93 per cent.

Infradiaphragmatic total anomalous pulmonary venous return is a rare form of congenital heart disease. Because of obstruction to pulmonary venous return these children present in the first few weeks of life with cyanosis, pulmonary oedema, and right heart failure. Medical management is unsuccessful; surgical treatment is difficult and there have been only sporadic successes.

It is the purpose of this review to present our experience with this malformation discussing the clinical, pathological, and surgical aspects, in the hope of defining guidelines for diagnosis and successful surgical management.

Subjects and methods

We have reviewed the clinical records, including data from cardiac catheterisation studies and, where indicated, the necropsy records of 28 patients with infradiaphragmatic total anomalous pulmonary

venous return seen at Texas Children's Hospital between 1957 and August 1975.

Cases of the mixed type of total anomalous pulmonary venous return were excluded from this review, as well as those with only partial anomalous venous return.

Cases were classified into two groups on the basis of associated intracardiac malformations. *Group A* (isolated infradiaphragmatic total anomalous pulmonary venous return) consisted of 17 patients with only a patent foramen ovale or atrial septal defect, and frequently a persistent ductus arteriosus. *Group B* (complex) consisted of 11 patients with infradiaphragmatic total anomalous pulmonary venous return who had other major intracardiac anomalies.

Ten cases in group B are discussed only from the viewpoint of the pathological anatomy and the association of splenic anomalies. The remaining case in this group is included in the review of cardiac catheterisation findings and surgical results. The clinical presentation, electrocardiograms, chest x-ray films, echocardiograms, cardiac catheterisation data, and surgical outcome of cases in group A

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Table 1 *Infradiaphragmatic total anomalous pulmonary venous return: anatomical findings*

	Group A Isolated	Group B Complex	Per cent of total
<i>Site of anomalous venous connection</i>			
Portal vein	11	8	68
Ductus venosus	2	0	7
Inferior vena cava	2	2	14
Left hepatic vein	1	1	7
Unknown	1	0	4
<i>Type of interatrial communication</i>			
Patent foramen ovale	14	5	68
Atrial septal defect	3	5	28
Common atrium	0	1	4
<i>Ductus arteriosus</i>			
Widely patent	9	1	36
Probe patent	2	1	11
Patent (unspecified size)	4	3	25
Closed	2	4	21
Unknown	0	2	7
Total	17	11	100

are presented in some detail.

Fourteen patients in group A had a cardiac catheterisation performed. Eleven of these and the case from group B had adequate haemodynamic records. The systemic arterial systolic pressure and oxygen saturation were compared with the pulmonary arterial systolic pressure and oxygen saturation in each case. Pulmonary hypertension was defined as a pulmonary arterial systolic pressure which was more than 50 per cent of systemic arterial systolic pressure. The pulmonary arterial oxygen saturation was considered equal to the systemic arterial oxygen saturation if there was no greater than two percentage points difference in oxygen saturation.

Eight patients in group A and 1 patient in group B had a surgical operation. Two patients from group A and the 1 from group B have had a postoperative cardiac catheterisation.

A necropsy was performed on 22 of the 25 patients who died. The development of left heart structures in patients from group A is further discussed in relation to the size of the interatrial communication and the ultimate surgical outcome. When describing the interatrial communication, use of the term atrial septal defect implies a larger opening than patent foramen ovale as evaluated at cardiac catheterisation, operation, or necropsy.

Results

SITES OF CONNECTION (Table 1)

In 19 cases (68%) the anomalous pulmonary venous connection was to the portal vein: this high frequency was similar in both groups A and B. Four

patients had connection to the inferior vena cava, 2 to the ductus venosus, and 2 to the left hepatic vein. In 1 case from group A, infradiaphragmatic total anomalous pulmonary venous return was seen angiographically but the exact site of connection was not defined; a necropsy was not obtained in this case.

INTERATRIAL COMMUNICATION (Table 1)

Fourteen cases from group A had a patent foramen ovale rather than an atrial septal defect. On the other hand, out of a total of 8 cases with an atrial septal defect 5 were from group B. One case in group B had a common atrium.

DUCTUS ARTERIOSUS (Table 1)

Twenty cases (72%) out of the total group had a persistent ductus arteriosus. In 10 of these the ductus was widely patent and in 3 probe patent. A further 7 cases had a persistent ductus arteriosus but its size was not described.

LEFT ATRIUM AND LEFT VENTRICLE (Table 2)

Eleven cases in group A with a patent foramen ovale had small left heart chambers; the left atrium especially was underdeveloped. Of 3 cases in group A with an atrial septal defect, 2 had a left atrium and left ventricle of normal size.

Table 2 *Size of the left heart chambers related to type of interatrial communication (Group A patients)*

	Reduced	Slightly reduced	Normal	Unknown
Patent foramen ovale	8	3	1	2
Atrial septal defect	0	1	2	0
Total	8	4	3	2

ASSOCIATION OF SPLENIC ANOMALIES (Table 3)

Six cases in group B had asplenia, 3 a normal spleen, and in 2 cases the splenic status was unknown. The associated cardiac malformations are listed in Table 3. In group A, comment was made on the spleen at necropsy in 8 cases; a bilobed spleen in one case was the only abnormality noted.

CLINICAL FINDINGS (Group A)

The male to female sex ratio was 3:1. The birth-weights ranged from 2.0 kg to 3.9 kg with a mean of 3.1 kg. Only 1 infant was born prematurely at 35 weeks of gestation. The infants developed symptoms between the first and eighth day of life, with an average age at the time of onset of symptoms of 3 days. There was a mean delay from the time of

Table 3 *Infradiaphragmatic total anomalous pulmonary venous return and complex congenital heart disease (Group B patients)*

Interatrial communication	11
Persistent ductus arteriosus	5
<i>Splenic status</i>	
<i>Asplenia</i>	6
Endocardial cushion defect	2
Transposition of great arteries	2
Pulmonary atresia	2
Mitral atresia	3
Ventricular septal defect	3
Truncus arteriosus	1
Inferior vena cava drainage to left atrium	1
Persistent left superior vena cava	3
Dextrocardia	2
<i>Normal spleen</i>	3
Endocardial cushion defect	1
Double outlet right ventricle	1
Mitral atresia	1
Infundibular pulmonary stenosis	1
Single A-V valve	1
Transposition of great arteries	1
<i>Unknown</i>	2
Single ventricle, dextrocardia	1
Ventricular septal defect, persistent ductus arteriosus	1
Total number of patients	11

onset of symptoms until admission to Texas Children's Hospital of 11 days with a range of 1 day to 7 weeks. A further mean delay of 6 days occurred between the time of admission and operation or death.

Feeding difficulty and slow weight gain were the most common presenting symptoms. Tachypnoea was the most constant sign. Mild to moderate cyanosis was present in all, but in 2 patients it was an inconstant feature and in only 1 was it described as being severe. Half of the patients had a grade 1-2/6 ejection systolic murmur best heard along the left sternal border. In 2 patients, a soft systolic murmur was heard only over the liver. The liver was enlarged 3.0 cm or more below the right costal margin in 13 patients, while in 4 the size of the liver was normal at the initial examination.

RADIOGRAPHS (Group A)

The cardiothoracic ratio was normal in 15 patients and increased in 2 patients. Thirteen patients had radiographic evidence of pulmonary oedema, 3 had normal lung fields, and in 1 case the pulmonary vascular markings were reduced.

ELECTROCARDIOGRAMS (Group A)

Sinus rhythm was present in all but 1 patient who had a wandering atrial pacemaker. Only 1 patient had right atrial enlargement. Right axis deviation of the QRS complex in the frontal plane was present in each case, ranging from +120° to +160°. Definite right ventricular hypertrophy was present in 12 of 14 available tracings with a qR pattern in the right chest leads in 6 of these. One tracing showed probable right ventricular hypertrophy. In 1 patient the electrocardiogram was interpreted as normal on the first day of life. A paucity of left ventricular forces was a striking abnormality in half of the patients.

ECHOCARDIOGRAMS (2 patients from group A and 1 patient from group B)

The left atrial and left ventricular dimensions were slightly reduced in two infants in the newborn period. The common pulmonary venous chamber was not identified and the right ventricular dimension was normal. A large right ventricle, paradoxical septal motion, and the common pulmonary venous trunk were noted in case B1 (Table 4).

CARDIAC CATHETERISATION (11 patients from group A and 1 patient from group B)

Systemic arterial oxygen desaturation was present in every case, the range of systemic arterial oxygen saturation for the group being 40 to 93 per cent. The mean systemic arterial oxygen saturation was 65 per cent in the group of 8 patients with only a patent foramen ovale and 80 per cent in those with an atrial septal defect. In 6 patients, 5 from group A

Table 4 *Clinical, haemodynamic, and necropsy data: correlation with surgical outcome*

	Age Days	Age Weeks	Weight (kg)	PAP/SAP × 100%	Systemic arterial oxygen saturation (%)	ASD or PFO	LA	LV	Anastomosis	Outcome
<i>Group A</i>										
1	—	1	2.66	> 100	54	PFO	VS	VS	Widely patent	D 1 day
2	6	—	3.5	—	—	PFO	S	S	Widely patent	D 1 day
3	12	—	2.75	> 100	63	PFO	VS	—	Widely patent	D 1 day
4	9	—	3.0	100	70	PFO	S	S	Widely patent	D 1 day
5	—	6	2.9	> 100	77	PFO	S	S	Stenotic	D 1 day
6	—	4	3.0	90	62	PFO	S	S	Stenotic	D 3 months
7	—	8	3.2	60	93	PFO	—	—	Widely patent	A 13½ years
8	—	4	4.3	> 100	70	ASD	N	N	Widely patent	A 3½ years
<i>Group B</i>										
1	10 yrs	—	20.0	100	85	ASD	N	N	Widely patent	A 2 years

PAP=pulmonary arterial pressure; SAP=systemic arterial pressure; ASD=atrial septal defect; PFO=patent foramen ovale; LA=left atrium; LV=left ventricle; VS=very small; S=small; N=normal size; D=died; A=alive.

Fig. 1 Comparison of pulmonary arterial systolic pressure with systemic arterial systolic pressure.

and the 1 patient from group B, the saturations were similar in all cardiac chambers and in both great arteries. The oxygen saturation in the pulmonary artery was lower than aortic saturation in 4 patients and higher in 1.

All patients had raised pulmonary arterial pressure (Fig. 1). In 7 patients the pulmonary arterial pressure was higher than the systemic arterial pressure, in 2 patients the pressures were equal, and in the remaining 2 patients the pulmonary arterial pressure was lower than the systemic arterial pressure.

Two patients, cases B1 and A7, had at least intermittently nonobstructive anomalous pulmonary venous drainage to the inferior vena cava and had pulmonary arterial pressures which were 100 per cent and 60 per cent of systemic, respectively. The systemic arterial oxygen saturation in these 2 cases was 85 per cent and 93 per cent, respectively. Both had successful operations.

SURGICAL CORRECTION (Table 4)

Eight patients in group A and 1 patient in group B had operations for correction of infradiaphragmatic total anomalous pulmonary venous return. Table 4 relates surgical outcome to the relevant clinical, haemodynamic, and anatomical features in these cases.

The average age of the patients in group A at the time of operation was 20 days, with a range of 6 days to 8 weeks. The patient in group B (B1), who survived operation, was 10 years of age at the time of diagnosis and surgical correction (Duff *et al.*, 1975). The average weight of the patients in group A was 3.2 kg (2.66 to 4.3 kg). Those who survived

operation in this group had an average weight of 3.5 kg.

Five patients died in the immediate postoperative period from low output failure. In 2 patients the patent foramen ovale was closed surgically and in 3 it was left open. At necropsy, the left atrium was small in each of these patients and very small in 2 patients. The left ventricle was also small in 3 patients, being hypoplastic in 1 patient.

Two patients died as a result of stenosis at the site of surgical anastomosis, one (A5) in the immediate postoperative period and the other (A6) 3 months after operation.

Three patients, 2 from group A and 1 from group B are alive and well, 13½ years (A7), 3½ years (A8), and 2 years (B1) from the time of surgical correction.

POSTOPERATIVE CARDIAC CATHETERISATION

The postoperative haemodynamic data in the 3 long-term survivors are shown in Table 5. All 3 are asymptomatic. Almost complete resolution of pulmonary hypertension has taken place and each has normal systemic arterial oxygen saturation.

The interatrial communication was closed at operation in 2 of the long-term survivors. In patient A8, the atrial septal defect was not closed and now

Fig. 2 Case A7. Angiogram shows a normal left atrium and left ventricle after injection of contrast medium into the main pulmonary artery. All pulmonary venous return is to the left atrium, with no residual infradiaphragmatic drainage. The pulmonary veins are mildly dilated.

at 3½ years of age he has a left-to-right shunt with a pulmonary to systemic flow ratio of 2.5:1. An angiogram (Fig. 2) on patient A7 13½ years after corrective operation showed a left atrium and left ventricle of normal size; the pulmonary veins, which are slightly distended, all drain to the left atrium.

Discussion

Total anomalous pulmonary venous return comprises 1 to 3 per cent of all congenital heart lesions (Keith *et al.*, 1967; Bharati and Lev, 1973). The anomalous connection and drainage is below the diaphragm in 10 to 20 per cent of these cases (Burroughs and Edwards, 1960). The frequency of drainage to different anatomical infradiaphragmatic sites in our series is similar to that reported by others, with 68 per cent connecting to the portal vein (Burroughs and Edwards, 1960; Bonham Carter *et al.*, 1969). Associated complex cardiac lesions have been noted in 20 to 30 per cent of patients with total anomalous pulmonary venous return (Burroughs and Edwards, 1960). When the connection is below the diaphragm the incidence of these anomalies is increased to approximately 50 per cent (Lucas and Schmidt, 1968). In the present series, 40 per cent of the cases had associated complex intracardiac anomalies.

Splenic anomalies, especially asplenia, are common in total anomalous pulmonary venous return when this is associated with complex cardiac lesions (Bharati and Lev, 1973; Delisle *et al.*, 1976). In the present series, two-thirds of the cases in group B had asplenia and one-third had a normal spleen. Anomalies of the spleen are rare in the absence of complex cardiac defects; a bilobed spleen in 1 case was the only anomaly noted in 8 group A patients.

Cardiac output and the development of the left atrium and left ventricle in isolated infradiaphragmatic total anomalous pulmonary venous return depend on the size of the interatrial communication (Burroughs and Edwards, 1960). Graham *et al.* (1972), using biplane cineangiography, showed that the volume of the left atrium was reduced in all patients with all types of total anomalous pulmonary venous return, but left ventricular volume was reduced in only one-third of their cases. Only 3 of their patients, however, were less than 2 months of age and the majority had nonobstructive total anomalous pulmonary venous return. With obstruction to pulmonary venous return and a patent foramen ovale one would expect the hypoplasia of the left atrium and the left ventricle to be exaggerated. The pathological observations in the present series and in that of Bharati and Lev

support this contention (Bharati and Lev, 1973).

The clinical presentation of total anomalous pulmonary venous return, irrespective of the site of connection, is determined chiefly by the presence or absence of obstruction to pulmonary venous drainage. When the connection is infradiaphragmatic some degree of obstruction to blood flow is almost always present. The causes of obstruction in this situation have been reviewed elsewhere (Burroughs and Edwards, 1960; Hastreiter *et al.*, 1962; Tynan *et al.*, 1974; Delisle *et al.*, 1976). Three of the patients in this series with isolated infradiaphragmatic total anomalous pulmonary venous return had at least intermittently nonobstructive drainage: 2 of these had connection to the inferior vena cava, and 1 to the portal vein with a large persistent ductus venosus. The clinical picture in the presence of obstruction is characteristic (Lucas and Schmidt, 1968). However, it is important to emphasise that cyanosis may not be prominent and may be intermittent. In addition, the liver may be of normal size and occasionally a murmur may be audible over it.

The chest x-ray usually supports or even suggests the diagnosis of obstructed pulmonary venous return with a diffuse hazy granular pattern of pulmonary oedema and normal heart size; however, the lung vascularity may occasionally appear normal or even reduced. This latter finding was noted in 1 of our patients and has also been described by Friedli *et al.* (1971). When pulmonary venous obstruction is severe, leading to pulmonary hypertension, the presence of a persistent ductus arteriosus allows right-to-left shunting which further reduces pulmonary blood flow producing normal or oligemic lung fields on x-ray. These infants are very cyanotic; the systemic arterial oxygen saturation in our patient was 40 per cent and in the reported patient of Friedli *et al.* 32 per cent. When obstruction is intermittent, there may be considerable variation in the lung vascular markings over a period of a few hours. Therefore, repeated observations are of considerable importance in evaluating these patients. Radiographic differentiation from neonatal pneumonia, hyaline membrane disease, aspiration syndrome, and pulmonary haemorrhage may be difficult at times (Johnson *et al.*, 1958). However, from a clinical standpoint, infants with these conditions usually show signs of improvement with increasing age, while those with obstructed total anomalous pulmonary venous return deteriorate.

The electrocardiogram was helpful in supporting the diagnosis and almost invariably showed right ventricular hypertrophy (Gathman and Nadas, 1970; Rudolph, 1974). A qR pattern in the right praecordial leads and reduced positive electrical forces in the left praecordial leads were noted in half

of our cases. Both these features were present in patients with pulmonary hypertension, but did not distinguish those with moderate from those with severe pulmonary hypertension.

Echocardiographic criteria for the diagnosis of total anomalous pulmonary venous return, especially the cardiac and supracardiac varieties, have been outlined by Paquet and Gutgesell (1975) from this institution. We did not find the echocardiogram to be diagnostic, however, in 2 infants with infradiaphragmatic total anomalous pulmonary venous return. In patient 1 from group B, who was 10 years of age, the common pulmonary venous chamber was clearly seen dorsal to the left atrium as described by Paquet and Gutgesell. Its value in the infant with infradiaphragmatic total anomalous pulmonary venous return is primarily to exclude other causes of cyanotic congenital heart disease, such as transposition of the great arteries and tetralogy complexes. However, the demonstration of a small left atrium and perhaps also small left ventricle lends support to the diagnosis of anomalous pulmonary venous return.

Those patients with lower systemic arterial saturation at cardiac catheterisation, especially when this is below 70 per cent, and with pressure at systemic level or above in the pulmonary artery are likely to be obstructive in type. This has also been noted by others (Gathman and Nadas, 1970; Delisle *et al.*, 1976). In determining the site of connection of the anomalous trunk, variation in oxygen saturation between the inferior vena cava and the rest of the heart was inaccurate and unreliable. In general, however, the infants were not catheterised through the umbilical vein which would have facilitated more direct sampling of pulmonary

venous blood from the portal vein (Sneed, 1972). Alternatively, manipulation of the catheter into the hepatic vein should be attempted in all instances when the catheterisation is from the femoral vein as a high oxygen saturation at that site would be diagnostic of anomalous return either to the hepatic or to the portal vein.

For precise diagnosis, angiography is essential. As suggested by Tynan *et al.* (1974), if the anomalous trunk can be entered directly through its site of connection, e.g. to the portal vein, then injection of contrast medium into the common channel gives excellent anatomical definition of the whole pulmonary venous tree. Failing this ideal, selective branch pulmonary artery injections are much more useful than a main pulmonary artery injection. From the latter site, in the presence of severe pulmonary hypertension, most of the contrast medium is lost through the persistent ductus arteriosus, which was patent in 82 per cent of isolated cases in our series. It is also important to continue the cineangiogram for 10 seconds or more to allow the contrast medium to circulate through the pulmonary vascular bed. An effort should be made to visualise all pulmonary veins, as there is often considerable variation in the site of attachment of individual veins to the common pulmonary venous channel. This has important surgical consequences as one or more pulmonary veins may be overlooked and continue to drain into the systemic venous circulation after distal ligation of the common channel (e.g. case B1, Table 5).

There is little argument that the proper management of isolated obstructed infradiaphragmatic total anomalous pulmonary venous return is surgical. The operation should be performed as an emergency as

Table 5 Postoperative cardiac catheterisation data

Patient	Site of drainage	Age	Haemodynamic data								Comment		
			Preoperative				Postoperative						
			Operation	Present	Oxygen saturation (%)		Pressure (mmHg)		Oxygen saturation (%)			Pressure (mmHg)	
					PA	Systemic A	PA	Systemic A	PA	Systemic A		PA	Systemic A
A7	IVC	8 wk	13½ yr	86	93	45/16	80/35	78	98	30/15	—	No residual defects	
A8	Portal vein	4 wk	3½ yr	63	70	100/50	80/50	78	96	28/12	100/0 10 (LV)	Residual atrial septal defect with Qp/Qs 2.5:1	
B1	IVC	10 yr	12 yr	85	85	110/58	110/80	76	98	45/11	105/76	Residual drainage of inferior left pulmonary vein to inferior vena cava but no step-up in oxygen saturation; mild tricuspid regurgitation	

Qp/Qs = ratio of pulmonary to systemic blood flow.

soon as the diagnosis is made, since these infants may deteriorate rapidly. There is an almost 100 per cent mortality with medical management, the majority of infants dying before 6 months of age (Keith *et al.*, 1967). In the presence of obstruction which is not at atrial level, balloon septostomy is not recommended at the time of cardiac catheterisation (El-Said *et al.*, 1972). Though surgical mortality is high and successes still worth reporting, excellent results can be achieved as shown by the 3 long-term survivors in this series. Including the patients in this review, 21 patients have had successful corrective operations for infradiaphragmatic total anomalous pulmonary venous return (Cooley and Balas, 1962; Sloan *et al.*, 1962; Woodwark *et al.*, 1963; Cooley *et al.*, 1966; Jeiger *et al.*, 1967; Llewellyn *et al.*, 1968; Mody *et al.*, 1969; Barratt-Boyes *et al.*, 1971; Friedli *et al.*, 1971; Joffe *et al.*, 1971; Buckley *et al.*, 1972; Barratt-Boyes, 1973; Breckenridge *et al.*, 1973; Higashino *et al.*, 1974). During follow-up, which has varied from 2 weeks to 13 years, 2 of these patients have died. The others were stated to be well at last evaluation, though only 2 have had haemodynamic evaluation postoperatively (Jeiger *et al.*, 1967; Higashino *et al.*, 1974).

The factors which contributed to the high mortality and militated against successful surgical outcome in the present series were:

- (1) Critically ill infants: this was often because of delay in clinical recognition of heart disease before hospital admission and delay in definitive diagnosis after admission.
- (2) A patent foramen ovale rather than an atrial septal defect: this was frequently associated with a small left atrium and left ventricle, which we felt contributed significantly to low output failure and death in 5 patients in the immediate postoperative period.
- (3) Systemic arterial oxygen saturation less than 70 per cent.
- (4) A pulmonary arterial pressure greater than systemic arterial pressure.

It seems also, both from the volume studies of Graham *et al.* and our subjective impression, that further reduction in the size of the small left atrium by primary closure of the intra-atrial communication is contraindicated.

Conclusion

With careful attention to clinical detail, critical radiographic evaluation, especially analysis of x-ray films taken at intervals of a few hours coinciding with alteration in clinical signs, supportive electrocardiographic and echocardiographic find-

ings, the diagnosis of total anomalous pulmonary venous return should at least be suspected. With this suspicion, cardiac catheterisation is performed to confirm the diagnosis and define the exact anatomical site or sites of connection and drainage. In this way, unnecessary delays can be avoided and operation performed when the infant is in the most stable cardiac and metabolic state.

Addendum

Since initially submitting this paper, we have had 3 additional newborn infants with isolated obstructive infradiaphragmatic total anomalous pulmonary venous return. One died while being prepared for operation and another died within 24 hours of surgical correction; the remaining patient had successful surgical correction at 5 days of age and is clinically well 2 months later.

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