Double-outlet right ventricle with intact ventricular septum

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In the present case report, we describe a malformation consisting of a double-outlet right ventricle with an intact ventricular septum diagnosed in a 3-day-old female newborn. To our knowledge 35 cases have been described in the literature. The diagnosis was made by two-dimensional echocardiography and confirmed by angiocardiography. An inadequate opening in the interatrial septum and hypoplasia of the mitral valve and left ventricle were present. In this condition the only outlet of the left ventricle is via the atrial septal defect. Balloon atrial septostomy was performed. At 1 month of age the patient required a right modified Blalock-Taussig shunt and surgical atrial septectomy. Five months after surgery, the patient was in good clinical conditions.

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The origin of both great vessels from the right ventricle with an intact ventricular septum is an extremely rare malformation described for the first time by MacMahon and Lipa[1]. In the 35 cases reported so far in the literature (Table I)[1-16], the anomaly was recognized at autopsy[1-3,5,8,10,11], angiography[9,12], and more recently at magnetic resonance imaging and fetal echocardiography[15].

In the present case report we describe a new case of this rare malformation identified in a 3-day-old female newborn and, for the first time, diagnosed by means of two-dimensional echocardiography.

Case report

The patient was delivered at term. Mild cyanosis was noted in the immediate postnatal period. The second heart sound was single. A 2/6 systolic ejection murmur was identified over the left sternal border. An electrocardiogram showed sinus tachycardia and right ventricular hypertrophy. Chest roentgenogram showed an enlarged heart; the lungs were clear. Two-dimensional echocardiographic study revealed *situs solitus* of the viscera and atria, levocardia, D-loop ventricles, a double-outlet right ventricle, an intact ventricular septum, malposition of the great arteries with the aorta strictly anterior to the pulmonary artery, bilateral conus, and severe pulmonary stenosis (Fig. 1). The mitral valve was stenotic, and a minor degree of hypoplasia of the left ventricular cavity was noted. The left ventricular outflow tract was atretic. The pulmonary venous return gained access to the right heart through a restricted atrial septal defect. There was a left-sided aortic arch with a left patent duct. Cardiac catheterization performed at 4 days of age showed no step-up in oxygen saturation from the right atrium to the right ventricle and aorta. The arterial oxygen saturation was 79%. The systolic pressure in the right ventricle was at systemic levels (50 mmHg) whereas the left ventricular pressure was above systemic values (75 mmHg). The mean pressure gradient between the left and right atria was 8 mmHg. Angiography confirmed the echocardiographic findings (Fig. 2). Balloon atrial septostomy enlarged the atrial septal defect from 3 to 6 mm and reduced the interatrial mean pressure gradient to 2-3 mmHg. At 1 month of age the patient required a right modified Blalock-Taussig shunt and surgical atrial septectomy because of progressive cyanosis. Five months after surgery, the patient was in good clinical conditions.

Table I. Cases reported in the literature.

Year/author	Age/sex	Diagnosis	MV	LV	ASD	Outflow stenosis
1964 MacMahon and Lipa[1]	3 months/F	Autopsy	MS, extreme	Hypopl marked	PFO	None
1965 Ainger[2]	2 years/M	Angio/autopsy	MS, extreme	Hypopl marked	Small	None
1968 Davachi et al.[3]	2 days/M	Autopsy	MS, extreme	Hypopl	Small	AS or hypoplasia
1968 Martin et al.[4]	6 days/M	Autopsy	MAt	(Cited by Descalzo)	Absent	
1972 Lev et al.[5]	*	Autopsy	MAt	"Grossly absent"	Small	PS
Lev	*	Autopsy	MAt	"Small"	Small	PS
Lev	**	Autopsy	MAt	Hypopl marked	Small	AS or hypoplasia
Lev	**	Autopsy	MAt	Hypopl marked	Small	AS or hypoplasia
Lev	**	Autopsy	MAt	Hypopl marked	Small	AS or hypoplasia
Lev	**	Autopsy	MS	Hypopl marked	Small	AS or hypoplasia
1972 Lunel et al.[6]	2 weeks/M	Autopsy	MS	Hypopl	Small	NK
1975 Zamora et al.[7]		Autopsy	Parachute MV	Hypopl	Present	AS
Zamora		Autopsy	MS	Hypopl	Present	AS or hypoplasia
1976 Cameron et al.[8]	4 days	Autopsy	MS, extreme	Hypopl	Absent	None
Cameron	4 days	Autopsy	MAt	Hypopl marked	Absent	AS
Cameron	1 month	Autopsy	MAt	Hypopl marked	Present	None
Cameron	3 months	Autopsy	MAt	Hypopl marked	Absent	PS
Cameron	5 months	Autopsy	MAt	Hypopl marked	Present	PS
Cameron	8 months	Autopsy	MAt	Hypopl marked	Absent	PS
1976 Sridaromont et al.[9]	5 years/F	Angio/intraop	Parachute MV	Hypopl	Present	None
Sridaromont	2 months/F	Angio/intraop	MAt	Hypopl	Present	None
1979 Descalzo et al.[10]	4 months/M	Angio/autopsy	MAt	Hypopl marked	PFO	PS
1982 Van Praagh et al.[11]	9 days/M	Autopsy	MAt	Hypopl marked	PFO	PS
Van Praagh	NK/M	Autopsy	MAt	Hypopl marked	Surgical	PS
Van Praagh	NK/NK	Autopsy	MS, extreme	Hypopl marked	Present	None
Van Praagh	14 days/M	Autopsy	MAt	Hypopl marked	Present	AS
Van Praagh	NK/M	Autopsy	MAt	Hypopl marked	PFO	PA
Van Praagh	42 hours/F	Autopsy	MS, extreme	Hypopl extreme	Absent	AS
Van Praagh	NK/M	Autopsy	MAt (right)§	Hypopl marked	Absent	PS
Van Praagh	3 years/F	Autopsy	MS, extreme§	Hypopl marked	Present	PS
1985 Accorsi and Thiene[12]	6 months/F	Angio	MS	Hypopl	Small	PS
1987 Pandit et al.[13]	(abstract not available)					
1997 Ikemoto et al.[14]	NK/M	MRI	MAt	Hypopl	Small	(Cited by Patel)
1999 Patel et al.[15]	21 weeks	Fetal echo	MAt	Absent cavity	Small	None/PS autopsy
2000 Cheung et al.[16]	Newborn/M	Echo+angio	MS	Hypopl	Small	PS
2001 Vairo et al. (this report)	3 days/F	Echo+angio	MS	Hypopl	Small	PS

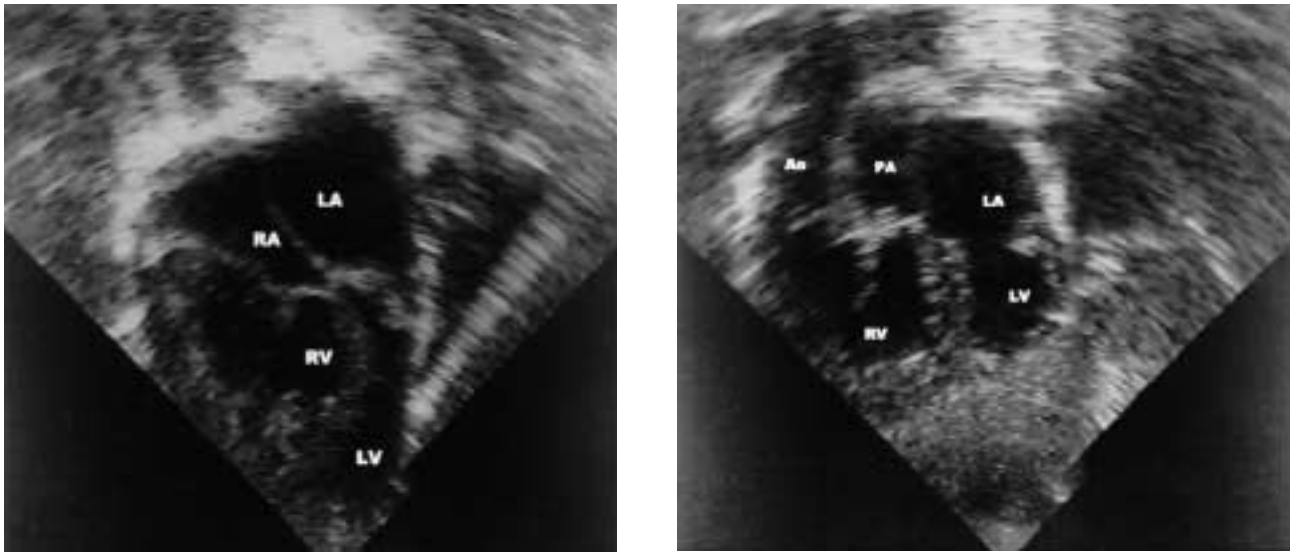
Angio = angiography; AS = aortic stenosis, ASD = atrial septal defect; Echo = echocardiography; Hypopl = hypoplastic; intraop = intraoperative; LV = left ventricle; MAt = mitral atresia; MRI = magnetic resonance imaging; MS = mitral stenosis; MV = mitral valve; NK = not known; PA = pulmonary atresia; PFO = patent foramen ovale; PS = pulmonary stenosis. * the age at death ranged from 15 days to 6 months with a mean of 2 months and 20 days; ** the age at death ranged from 1 day to 10 weeks, with a mean of 22 days; § situs solitus, L-loop ventricles, L-malposition of the great arteries.

Discussion

The absence of a ventricular septal defect in a double-outlet right ventricle is a rarity. Pathogenetic hypotheses included spontaneous closure of the ventricular septal defect and defective conal absorption[12]. A crucial problem in this condition is the presence of an inadequate channel for the flow of blood from the left side of the heart. Blood flow is thus shunted through an opening in the atrial septum[3].

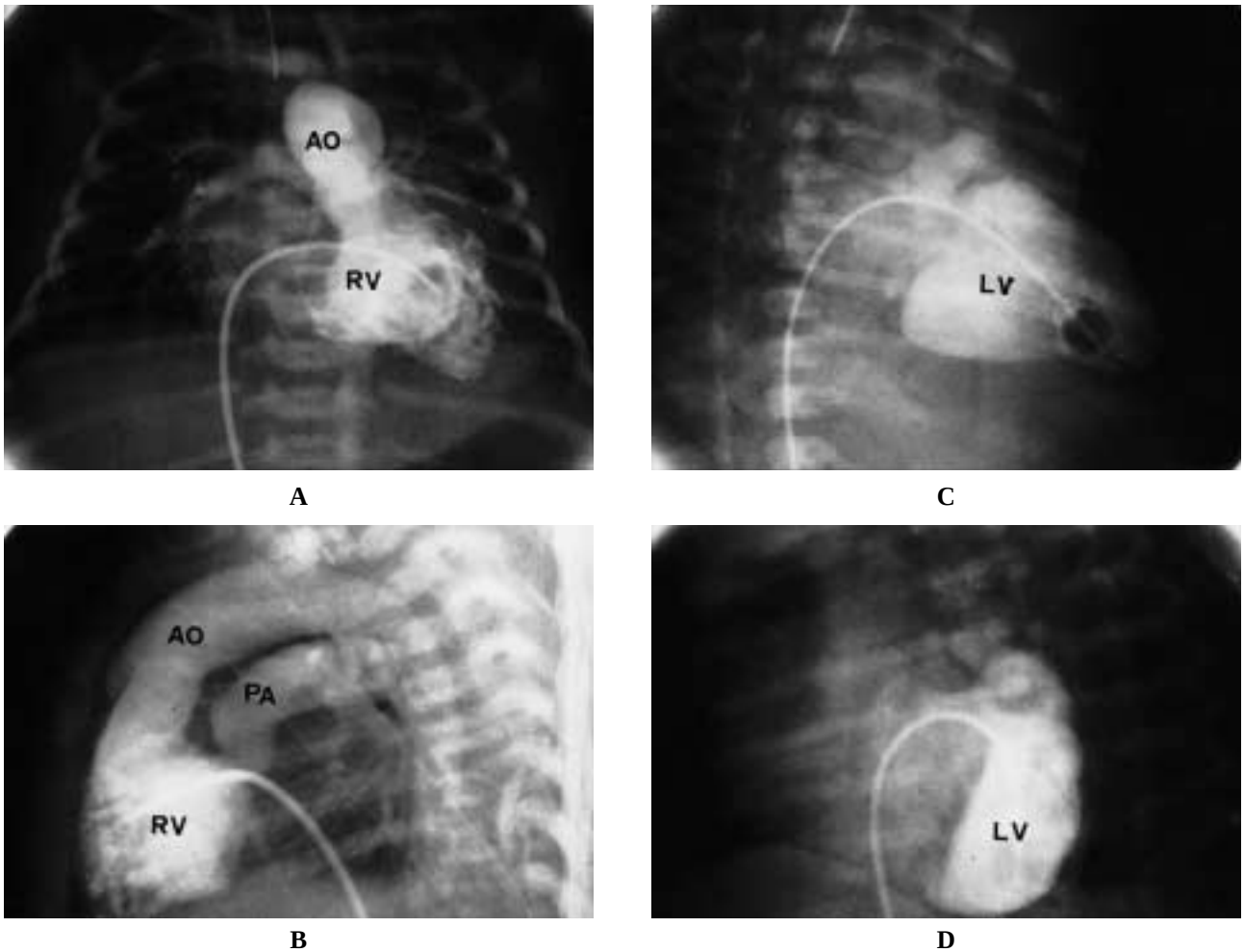
The patient with an intact ventricular septum usually presents with *situs solitus*, D-loop ventricles and malposition of the great arteries (D-malposition, side-by-side or A-malposition of great arteries). Only 2 cases of such malformation have been reported by Van Praagh et al.[11], with L-loop ventricles and L-malposition of great arteries. An atrial septal defect and a hypoplastic mitral valve (mitral stenosis or mitral atresia) and left ventricle were constantly present. Van Praagh et al.[11] described 8 cases of a double-outlet right ventricle without a ventricular septal defect, 4 including a subpulmonary conus and 4 a bilateral conus. Occasionally, however, a subaortic conus has been reported. Pulmonary stenosis occurs in a considerable number of cases. The absence of outflow tract obstruction was occasionally observed (Table I). Aortic stenosis is usually associated with infundibular stenosis, a hypoplastic ascending aorta, a hypoplastic aortic arch and aortic coarctation. The aortic arch is frequently left-sided.

In our case report, differently from all previously described cases, the mitral valve and left ventricle were only mildly hypoplastic.



A B

Figure 1. *Cross-sectional echocardiographic examination.* A: *apical 4-chamber view. An atrial septal defect and a hypoplastic mitral valve and left ventricle were present.* B: *left oblique subxiphoid view: the great arteries arise from the right ventricle with both arterial valves at the same level with the muscular outlet septum between them. Ao = aorta; LA = left atrium; LV = left ventricle; PA = pulmonary artery; RA = right atrium; RV = right ventricle.*



A C

B D

Figure 2. *Angiographic study.* A *and* B, *frontal and lateral views of a right ventricular angiocardiogram demonstrating simultaneous opacification of the aorta and pulmonary artery. The aorta is straight anterior (A-malposition of the great arteries). The aortic arch is left-sided;* C *and* D, *right and left oblique views of a left ventricular angiocardiogram (after left atrial decompression), demonstrating stenosis of the mitral valve, moderate hypoplasia of the left ventricle and an intact ventricular septum. Abbreviations as in figure 1.*

The impaired right ventricular function would constitute a setback if a total cavopulmonary connection or by other modifications of the Fontan principle were to be employed[16].

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