Virchows Arch. A Path. Anat. and Histol. 388, 39–49 (1980)

Virchows Archiv A
© by Springer-Verlag 1980

Double Inlet Right Ventricle

Report of a Case and Review of the Literature

J.N. Cox, P. Bopp, and E. Hauf

Departments of Pathology and Medicine (Division of Cardiology) and Cardiovascular Surgery, Geneva, Switzerland

Summary. A case of double inlet right ventricle in which both atrioventricular valves opened completely into the right ventricle is described. The left atrium communicated with the right ventricle by way of an opening low in the atrial septum and there was a large ventricular septal defect. The pulmonary trunk and aorta took their origin from the right and hypoplastic left ventricles respectively. The literature on this rare cardiac anomaly is reviewed and the morphological findings in this case are compared with those of the recorded cases. A short discussion of the possible embryogenesis is given.

Key words: Double inlet right ventricle – Straddling mitral valve – Double outlet right ventricle – Common ventricle.

Introduction

Double inlet right ventricle is a very uncommon abnormality which is characterized by the partial or total opening of both atrioventricular valves into the right ventricle. This condition has often been described in association with other cardiac malformations. The exact number of cases described in the literature is uncertain and depends largely on the authors' definition of this anomaly. Using the criteria of Tendon et al. (1973) only six such cases would classify as double inlet right ventricle, while those of Quero-Jimenez et al. (1973) would bring this number to eleven. All the cases thus far reported presented with double outlet right ventricle among other anomalies. The present case is different in a number of respects, including the origin of each great vessel from a separate conus and the long survival.

Offprint requests to: J.N. Cox, M.D., Department of Pathology, 40, Boulevard de la Cluse, CH-1211 Geneva 4, Switzerland

0340-1227/80/0388/0039/$02.20

Case Report

Clinical Features

This 16 year-old Moroccan boy was born (1957) after an uneventful pregnancy. He was cyanosed at birth, and this became accentuated with time. During childhood dyspnoea was noticed on exertion, but there were no fainting spells. Physical growth was somewhat retarded but mental development was considered normal.

The boy weighed 38 kg for a height of 157 cm. There was marked cyanosis of the extremities and clubbing of the fingers. Moderate bulging of the left side of the chest-wall was noted but there was no cervical venous distension. The pulse rate was regular at 90/min; all pulses were easily palpated; blood pressure was 120/80 mm Hg. The edge of the liver was at the costal margin and there was no peripheral oedema.

On auscultation there was a right ventricular heave and loud heart sounds with a single second sound over the pulmonary area. A grade III/IV pansystolic murmur was audible at the second left intercostal space.

Laboratory examinations indicated a severe polycythaemia of 9,200,000 RBC/mm^3, Htc at 76% and Hb of 20.3 g%. The relevant biochemical and other examinations were within normal limits.

A chest X-ray showed moderate enlargement of the right ventricle and straightening of the cardiac contour on the left side. The electrocardiogram showed regular sinus rhythm, a right axis deviation (+120°) with right auricular and ventricular hypertrophic patterns.

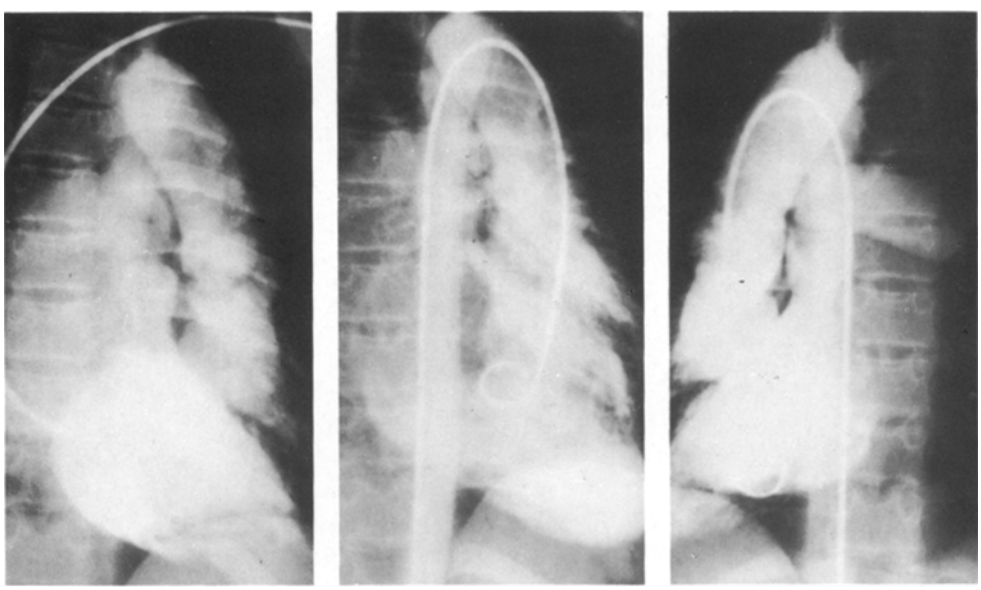


Fig. 1a–c. Angiocardiograms. **a** Lateral view of right ventricular angiocardiogram showing the dilated, coarsely trabeculated right ventricle, the stenosed pulmonary valvular ring. There is simultaneous filling of the hypoplastic left ventricle and the aorta situated in front and above the right ventricle. **b** Lateral view of left side catheterization showing the catheter crossing the hypoplastic left ventricle with its poorly trabeculated wall and entering the right ventricle by way of the ventricular septal defect. The pulmonary trunk and its stenosed valvular ring are visible. **c** Left lateral view in which the hypoplastic left ventricle and aorta are situated in front and above the dilated right ventricle and pulmonary trunk. The stenosed pulmonary valve is on the same plane with the aortic valve

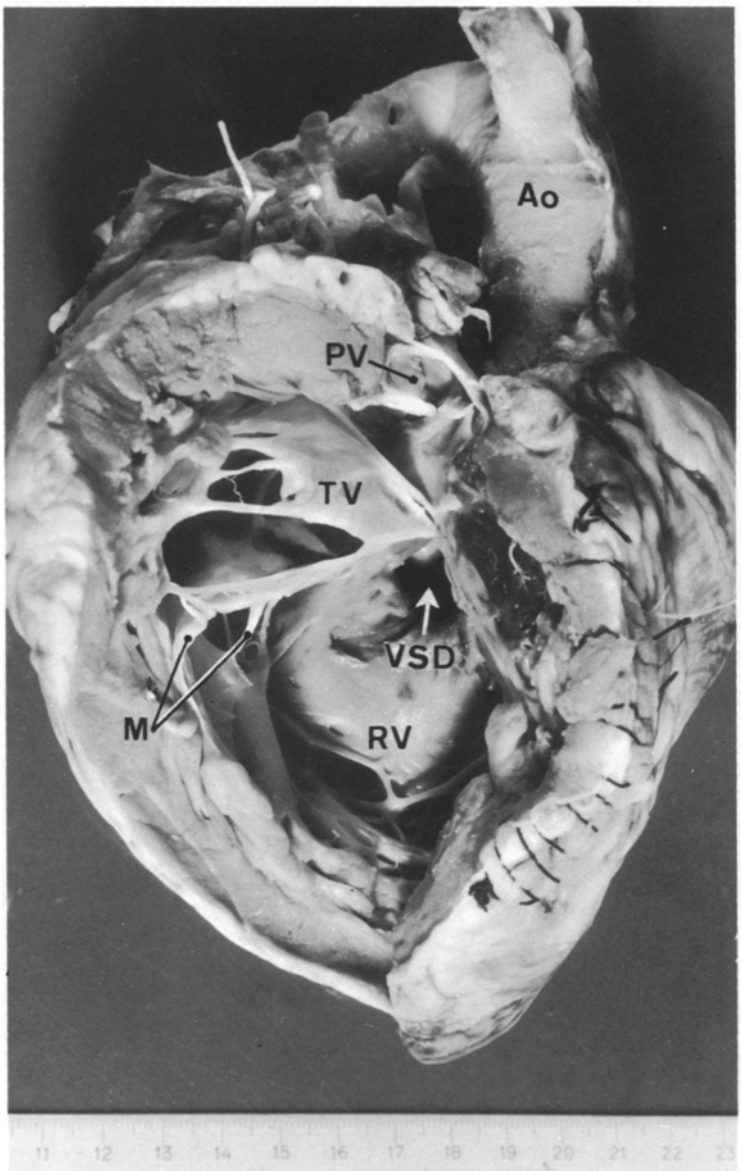


Fig. 2. The right ventricle (*RV*) is opened showing the large tricuspid valve (*TV*) in front and above the smaller mitral valve (*M*). The anterior leaflet of the tricuspid valve forms part of the floor of the infundibulum and is in close relationship with the stenosed pulmonary valve (*PV*). A_o aorta, *VSD* ventricular septal defect

At cardiac catheterisation systolic pressures were balanced in the left and right ventricle and there was a gradient of 90 mm Hg at the pulmonary valvular level, but none at the aortic and subaortic levels. Oxymetry values were compatible with bidirectional intracardiac shunts. An angiocardiogram (right ventricular injection by means of a catheter introduced into the right cubital vein), in the antero-posterior incidence, showed good filling of an enlarged coarsely trabeculated

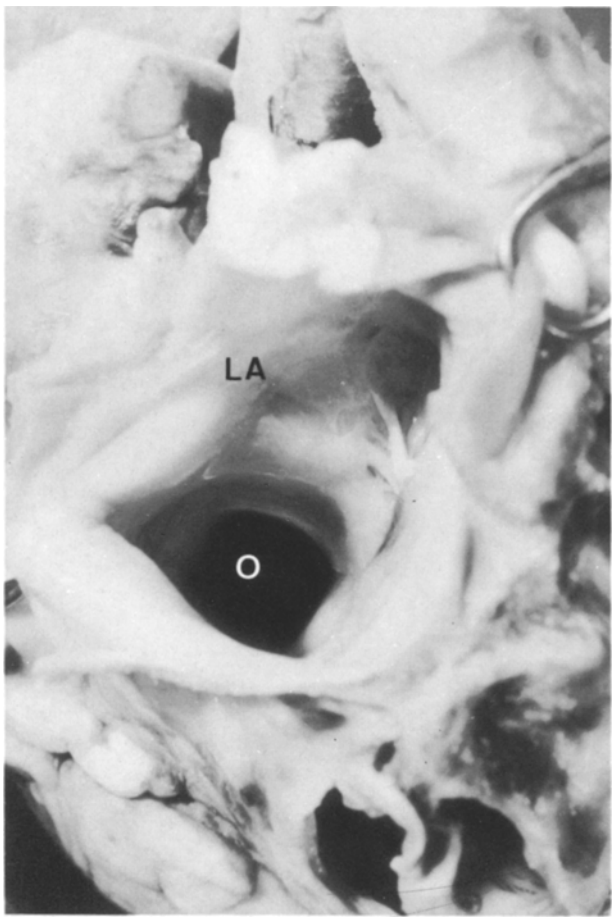


Fig. 3. View from above of the left atrium showing the orifice (*O*) low in the atrial septum and opening in the right ventricle

right ventricle with simultaneous opacification of both the pulmonary trunk and aorta which were on the same plane in the lateral view (Fig. 1a). The pulmonary valve appeared thickened with a dome-like systolic image. On subsequent films there was opacification of the right atrium immediately after that of the left atrium. On left side catheterization by way of the aorta, the catheter attained the right ventricular cavity through a high ventricular septal defect. The hypoplastic left ventricle in front of the dilated right ventricle was visualized again simultaneously (Figs. 1b, 1c). No selective left atriogram was performed. A retrograde arterial ventriculogram confirmed the hypoplastic left ventricle and showed a large anterior coronary artery.

The diagnosis of double outlet right ventricle with pulmonary valvular stenosis, malposition of the great vessels, atrial and ventricular septal defects and hypoplasia of the left ventricle was made. Surgery was undertaken to correct this condition, but the patient succombed shortly after the intervention.

Pathologic Findings

A 812/73: The configuration of the organs was that of situs solitus. The major pathological findings were limited to the *heart*. The heart was enlarged, weighing

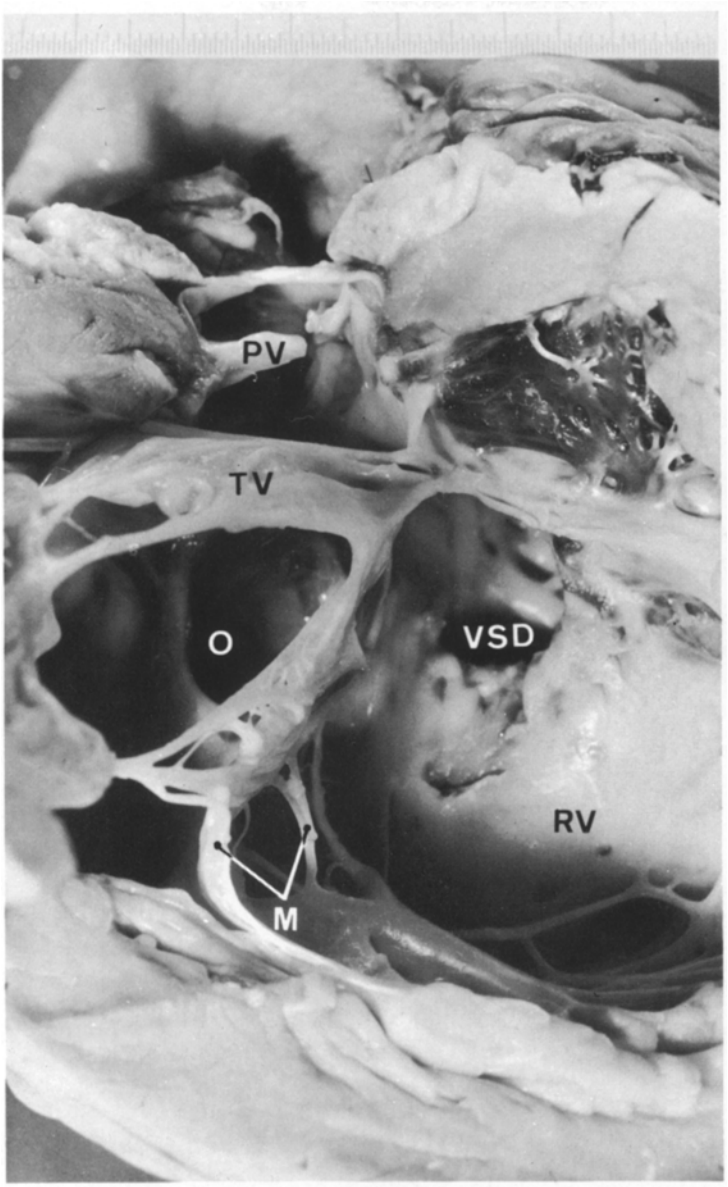


Fig. 4. Detail view of the right ventricle (*RV*) and the atrioventricular canal. The orifice (*O*) in the atrial septum is clearly visible, with the anterior leaflet of the tricuspid valve (*TV*) situated above while its septal leaflet whose edge is fused with the anterior leaflet of the mitral valve (*M*) over-rides this orifice (*O*). *PV* pulmonary valve; *VSD* ventricular septal defect

350 g, and its apex was formed by the right ventricle which was directed to the left.

The anatomically *right atrium*, hypertrophied and dilated, was in its normal position. It received the *superior vena cava* and the dilated *inferior vena cava*, both in their normal position. The foramen ovale was closed and, like the

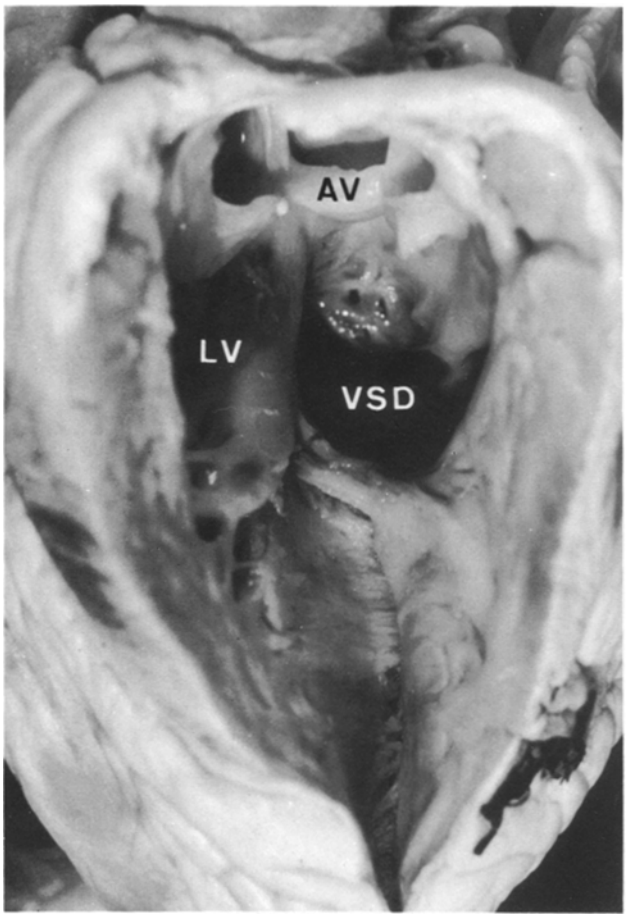
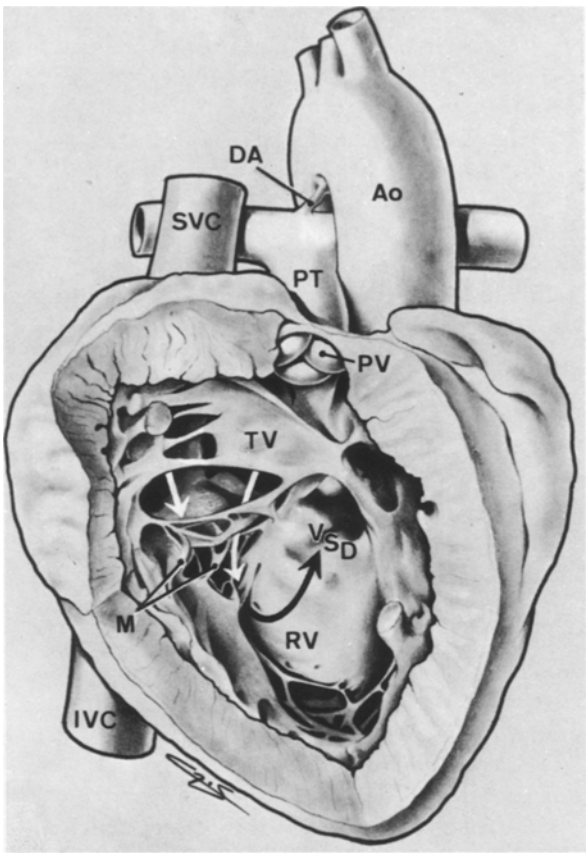


Fig. 5. View of the hypoplastic left ventricle (*LV*) showing its thickened wall with the few, small trabecules. *AV* aortic valve; *VSD* ventricular septal defect

coronary sinus, was normal. Both Eustachian and Thebesian valves were present but rudimentary. The right atrial appendage was dilated, thickened and free of thrombi.

The *right ventricle* was dilated, hypertrophied (2 to 2.4 cm thick) and markedly trabeculated. The right atrium opened into the right ventricle by way of the tricuspid valve (3.8 cm in diameter). The three leaflets of the tricuspid valve were large and their chordae, with the exception of those of the posterior cusp, were long and thin (Fig. 2). The septal cusp was somewhat fenestrated (five small orifices) near its anterior insertion.

The *pulmonary trunk* took its origin from the right ventricle. It was situated to the right and somewhat behind the aorta with little conus tissue. The anterior cusp of the tricuspid valve formed part of the floor of the infundibulum of the outflow track which it partially obliterated. The pulmonary orifice was stenosed (1.2 cm in diameter) but possessed three semi-lunar cusps, one of which (posterior) was small, thickened, retracted and rigid.



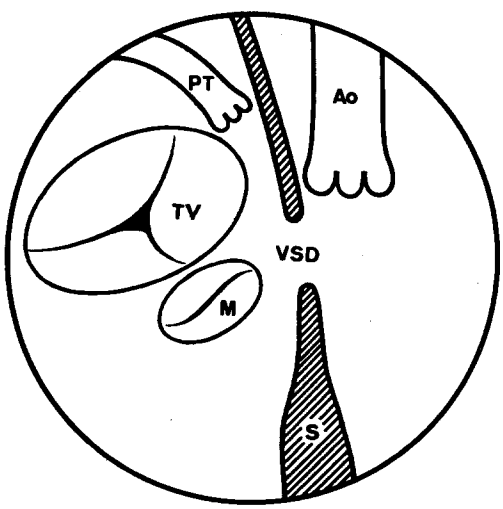


Fig. 6. a Schematic illustration of the direction of blood flow (*arrows*). **b** Diagram illustrating the position of the atrioventricular valves, pulmonary trunk and aorta. *SVC* superior vena cava; *IVC* inferior vena cava; *RV* right ventricle; *TV* tricuspid valve; *M* mitral valve; *PT* pulmonary trunk; *PV* pulmonary valve; *VSD* ventricular septal defect; *S* ventricular septum; A_o aorta; *DA* ductus arteriosus

The anatomically *left atrium* and its appendage were in their normal position but hypoplastic. The *pulmonary veins* emptied into the left atrium normally.

The *interatrial septum* pressented in its lower portion an oval orifice, 1.6 cm in diameter, which opened just above the atrioventricular ring, superior to the anterior and septal cusps of the tricuspid valve (Fig. 3). The septal cusp also presented an orifice, 1.2 cm in diameter, in its posterior portion.

There was no left atrioventricular canal between the left atrium and left ventricle. The left atrium was in communication rather with the right ventricle by the above orifice.

Behind the septal cusp of the tricuspid valve were the mitral valves, much smaller than normal (Fig. 4). The edge of the anterior cusp of the mitral valve was fused with that of the septal tricuspid leaflet and together they formed a bridge over-riding the interatrial orifice in the middle. The chordae were thin, short and were anchored to the right portion of the posterior papillary muscle. The smaller posterior cusp was fixed near to the upper portion of the atrioventricular septum. Its chordae were long, thin and were inserted on the thick portion of the posterior papillary muscle.

The *left ventricular cavity* was small (Fig. 5) with few trabeculae carneae. Its wall was hypertrophied (2.3 cm thick) and no papillary muscles were present. The cavity communicated with the *right ventricle* by way of an interventricular septal defect, 2 cm in diameter.

The *aorta* took its origin from the hypoplastic left ventricle. It was situated in front of the pulmonary trunk (D-loop). The aortic valves were normal but somewhat rotated to the right. The *coronary arteries* took their origin normally. The arterial branches from the aortic arch were normal. The ductus arteriosus was patent and measured 2.3 cm in length.

The final anatomical diagnosis was that of a double *inlet right ventricle* with both atrioventricular valves opening into the hypertrophied and dilated right ventricle, pulmonary stenosis and a hypoplastic left ventricle with an interventricular septal defect (Fig. 6). The aorta took its origin from the left ventricle and the ductus arteriosus was patent.

Discussion

Munoz-Castellanos et al. (1969) introduced the notion of double inlet right ventricle to describe, in a 3 year-old girl, a congenital cardiac abnormality in which both atrioventricular valves opened completely into a large hypertrophied anatomical right ventricle (double inlet), both great vessels taking their origin from this ventricle (double outlet); the left ventricle being present but hypoplastic.

A second case (girl, 6 years) was described by Munoz-Castellanos et al. (1973) and compared with the first. These cases differred in that the second heart presented a patent foramen ovale and no ventricular septal defect. These authors therefore considered double inlet right ventricle as an alteration in the normal development of the atrioventricular canal and its connections at an early stage of embryogenesis and thought that this group should be considered a separate entity distinct from the common ventricle.

In the case presented here both tricuspid and mitral valves were situated completely in the dilated, hypertrophied anatomical right ventricle. The right atrium was normally connected to the right ventricle while the mitral valve, placed to the left and somewhat behind the septal cusp, was also entirely within the right ventricle. It communicated with the left atrium by way of the opening in the lower portion of the interatrial septum. This orifice was partially divided by the somewhat fused leaflets of the septal tricuspid cusp and the anterior mitral cusp. There was no communication between the left atrium and the anatomic left ventricle from which the aorta took its origin. This abnormality to some extent would fulfill the theoretical hypothesis of Munoz-Castellanos et al. (1973) that both atrioventricular ostia may open completely into the right ventricle.

In 1973, Quero-Jimenez et al. described seven infants with congenital cardiac malformations, all of whom presented with a rightward displacement of the mitral valve into the right ventricle. Six of these were studied at post-mortem and they all showed varying degrees of displacement, from minor to major abnormalities.

Cases 1 and 2, comprising group A of this series, present some features common to the case under discussion. In both cases the tricuspid and mitral valves were contained in the morphologically right ventricles, although in Case 1 there was a common atrioventricular valve with a small mitral cusp over-riding the small ventricular septal defect. There was also an L-malposition of the great arteries with a bilateral conus. In the second case, there were two well developed valves and no ventricular septal defect but a double outlet right ventricle. The remaining four autopsied cases presented various degrees of a straddling mitral valve opening largely into the right ventricle.

An additional case was reported by Tandon et al. (1973) in which there was a straddling mitral valve communicating about equally with each ventricle. Double outlet right ventricle was an associated feature. Tandon et al. (1973), drawing certain parallelism with the more common condition known as double inlet left ventricle, in which the tricuspid valve straddles the ventricular septum onto the left ventricle, considered their case in which the mitral valve straddled the ventricular septum onto the right ventricle as being in the group called double inlet right ventricle. These authors considered only Cases 6 and 7 reported by Quero-Jimenez et al. (1973) and that of Munoz-Castellanos et al. (1969) as fulfilling the necessary criteria to which the term double inlet right ventricle could be applied.

A further case was later documented by Cabrera and Azcuna (1974) in a baby girl of 24 h, which resembled in many respects that of Case 2 (group B) of Quero-Jimenez et al. (1973) except that a small ventricular septal defect was present but was partially obliterated by the posterior cusp of the hypoplastic mitral valve.

From these various observations *double inlet right ventricle is characterized by the partial or total opening of both atrioventricular valves into the right ventricle;* the malposition of the mitral valve may vary from that of a straddling valve to one of complete displacement. The left ventricle is always present even though it may be markedly underdeveloped, and a ventricular septal defect may or

may not be present. Extreme displacements of the mitral valve have been referred to as a single right ventricle (Type B) of Van Praagh et al. (1964) (Quero-Jimenez et al. 1973) but, in this condition, the left ventricular sinus is absent.

Double inlet right ventricle must be distinguished from those cases in which there is mitral or tricuspid atresia, atrioventricular canal defects or the presence of accessory valves. This cardiac anomaly is closely related to double inlet left ventricle (Lev et al. 1969; Liberthson et al. 1971; Marin-Garcia et al. 1974; Van Praagh et al. 1964) as both conditions appear to develop early during embryogenesis when there might be an exaggerated displacement of the atrioventricular canal towards the bulbus cordis resulting in a rightward displacement of the mitral valve (double inlet right ventricle) or an arrest at the developmental stage when both atria are connected to the left ventricle (double inlet left ventricle) (Quero-Jimenez et al. 1973). Tandon et al. (1973) have further pointed out that there might be a malposition of the ventricular septum which could be associated with an abnormal direction of growth of the ventricular cavities with subsequent abnormal attachments of the atrioventricular valve to both sides of the septum. Another feature of the condition is the lack of alignment on the same plane of the interatrial and interventricular septa resulting in a ventricular septal defect.

In the case described in this report, the abnormality may have occurred at an early phase of development during the period when there is partitioning of the atrium, active proliferation of the endocardial cushions and closing of the interatrial foramen primum with separation of the atrioventricular canal, that is between the XIII and XVII Horizon of Streeter (Chuaqui and Bersch 1972). At this critical period there might have been an arrest in the final closing of the foramen primum with abnormal development of the ventral endocardial cushion and displacement of the atrioventricular canal towards the bulbus cordis. This would be in accordance with Doerr's theory on the morphogenesis of arterial transposition as the result of vectorial bulbus rotation and the existence of a teratological series, or spectrum, of anomalies pathogenetically related to transposition (Chuaqui 1979).

All cases of double inlet right ventricle so far documented have been reported in infants or young children. The case reported here has had the longest period of survival (16 years). Complete haemodynamic and angiocardiographic studies were only available in a few cases and even then the diagnosis had eluded the examiners. The clinical features, the electrocardiogram, the roentgenography and haemodynamic findings are all non specific. Quero-Jimenez et al. (1973) have suggested that selective left atrial angiography may yield clues to the diagnosis by showing a double stream of dye into the ventricular chambers. In addition, other anomalies such as double outlet right ventricle, transposition of the great vessels, ventricular septal defect are often associated, further complicating the general picture.

Surgical correction is clearly difficult and is limited to palliative procedures.

Acknowledgements. The authors wish to thank Mrs. C.L. de Marignac for secretarial assistance, Mssers J.C. Rumbeli and E. Denkinger for the photographs and Mr. G. Sesenna for the artistic drawings.

References

Cabrera A, Azcuna JI (1974) Ventricule droit à double entrée: étude d'un cas anatomo-clinique. Arch Mal Coeur 67:65–71

Chuaqui-JB, Bersch W (1972) The periods of determination of cardiac malformations. Virch Arch Abt A Path Anat 356:95–110

Chuaqui B (1979) Doerr's theory of morphogenesis of arterial transposition in light of recent research. Br Heart J 41:481–485

Lev M, Liberthson RR, Kirkpatrick JR, Eckner FAO, Arcilla RA (1969) Single (primitive) ventricle. Circulation 39:577–591

Liberthson RR, Paul MH, Muster AJ, Arcilla RA, Eckner FAO, Lev M (1971) Straddling and displaced atrioventricular orifices and valves with primitive ventricles. Circulation 43:213–226

Marin-Garcia J, Tandon R, Moller JH, Edwards JE (1974) Single ventricle with transposition. Circulation 49:994–1004

Munoz-Castellanos L, Rodriguez-Llorian A, Martinez-Rios MA and Espino Vela J (1969) Doble camara de salida y de entrada del ventriculo derecho. Arch Inst Cardiol Mexico 39:114–123

Munoz-Castellanos L, de la Cruz M, Cieslinski A (1973) Double inlet right ventricle: Two pathological specimens with comments on embryology. Br Heart J 35:292–297

Quero-Jimenez M, Perez-Martinez VM, Maitre Azcarate MJ, Merino-Batres G, Moreno-Granados F (1973) Exaggerated displacement of the atrioventricular canal towards the bulbus cordis (rightward displacement of the mitral valve). Br Heart J 35:65–74

Tandon R, Holler JH, Edwards JE (1973) Communication of mitral valve with both ventricles associated with double outlet right ventricle. Circulation 48:904–908

Van Praagh R, Ongley PA, Swan HJC (1964) Anatomic types of single or common ventricle in man. Morphologic and geometric aspects of 60 necropsied cases. Am J Cardiol 13:367–386

Accepted May 5, 1980