

Transatrial repair of double-outlet right ventricle in infants

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ABSTRACT In three infant cases of double outlet right ventricle (DORV), two with normally related great arteries (NGA) and one with side-by-side great arteries, a transatrial repair was carried out. In all three cases, the results were excellent. It is concluded that in the small baby with DORV with NGA and in DORV with side-by-side great arteries with a hypoplastic crista, a transatrial repair should be successful. This is dependent on the VSD being in the perimembranous (and, therefore, subaortic) location and on the absence of infundibular pulmonary stenosis. In all other varieties of DORV the repair should probably be done through the ventricle.

The repair of a double outlet right ventricle (DORV) is usually performed via a ventriculotomy.¹⁻⁵

Ionescu *et al*,⁶ however, reported two cases of DORV without pulmonic stenosis, in which the VSD was repaired via the right atrium. The atrial approach to closure of VSD became a common practice in recent years for the ordinary VSD in babies⁷⁻¹⁰ and we, too, prefer the right atrial approach to closure of a VSD. Recently we had the opportunity to use the transatrial approach for

repair of DORV in three infant cases and the purpose of this report is to analyse the anatomical features which allow the transatrial approach.

Case reports

CASE 1

A 23-month-old girl initially followed-up for suspected VSD and pulmonary hypertension in another hospital, was admitted to our medical centre where a diagnosis of DORV with NGA and pulmonary hypertension was made (table, fig 1). At operation a transatrial approach was used. The anterior tri-

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Table Haemodynamic and operative data

Haemodynamic data	Case 1		Case 2		Case 3	
	Preop	Postop	Preop	Postop	Preop	Postop
RV pressure (mm Hg)	120/0-13		110/0-10	70/0-5	75/0-10	60/0-5
MPA pressure	80/40		70/35	70/30	75/40	60/30
RPA pressure	30/20		70/35	70/30	75/40	
LV pressure		100/0-7	100/0-10			
Aortic pressure		100/50				
RV O ₂ Sat (%)	84		81	74.5	77	74
MPA O ₂ Sat (%)			80.5	75	80	73
Systemic O ₂ Sat (%)		95	91	95		95
Operative data						
Body temperature at surgery	24°C		25°C		27°C	
Total bypass time	58 min		59 min		68 min	
Total aortic cross-clamp	38 min		37 min		37 min	
Surgical result	Good		Good (residual pulmonary hypertension)		Good	

RV = right ventricle; MPA = main pulmonary artery; RPA = right pulmonary artery; LV = left ventricle.

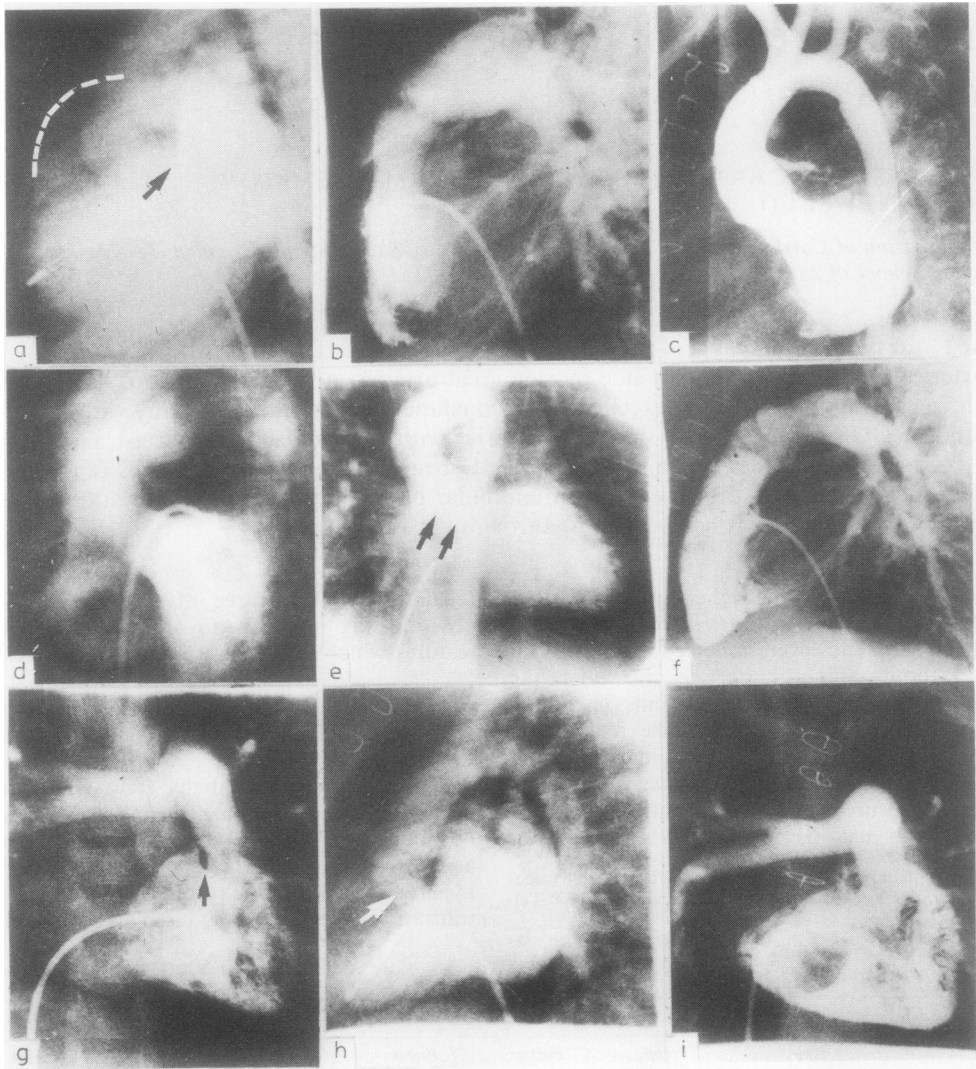


Fig 1 Pre- and postoperative angiograms of the three cases of DORV. (a-c—case 1; d-f—case 2; g-i—case 3). (a) Preoperative lateral view of case 1 showing the normal aortico-pulmonary relationships with persistent subaortic conus (black arrow). The white broken line demarcates the ill-defined border of the pulmonary artery. (b) Same as (a) demonstrating the pulmonary artery. (c) Postoperative lateral view of case 1, showing the large left ventricular to aortic tunnel. (d) Preoperative left anterior oblique angiogram of case 2, showing both great arteries arising from the right ventricle with normal semilunar interrelationships. (Because of the oblique exposure, the two arteries are in the same plane.) (e) Postoperative angiogram of case 2 showing, by means of recirculation, a large left ventricular to aortic tunnel (two black arrows). (f) Postoperative lateral view of the right ventricular outflow tract in case 2. Notice the normal anterior location of the pulmonary artery. (g) Preoperative AP angiogram of case 3, showing the hypoplastic crista (black arrow) and typical side-by-side great arteries. (h) Postoperative lateral angiogram of case 3 showing, in recirculation, that the left ventricular to aortic tunnel (white arrow) is of a good size. (i) Postoperative AP angiogram showing the RV outflow tract of case 3.

cuspid leaflet was incised along its base. Via the incision in the anterior tricuspid leaflet a Teflon felt patch was sutured as a conduit from the VSD to the aorta. In the region of the parietal muscle band which interrupts the aortico-tricuspid fibrous continuity (that is the subaortic conus), the Teflon patch was sutured along the tricuspid annulus. This leaves the aortic conus inside the left ventricular-aortic tunnel. (By so doing, the risk of creating subaortic stenosis is practically eliminated.) The patient was discharged on the seventh day after operation and readmitted six months later for recatheterisation. She was not taking any medications and her clinical condition was good. Because of technical problems only left heart catheterisation was performed and revealed excellent operative results (table, fig 1).

CASE 2

The clinical history, catheterisation data, operative findings, and immediate postoperative results were identical to those of case 1. Recatheterisation was done four months after operation and revealed normal function of the right ventricle and persisting moderate pulmonary hypertension. Left heart study was not performed but recirculation revealed maximal size of the LV outflow tract (table, fig 1).

CASE 3

A 15-month-old girl with the clinical picture of VSD and pulmonary hypertension was admitted for cardiac catheterisation and operation. A diagnosis of classical DORV with small crista and great arteries situated side by side was made (table, fig 1). At operation, through the right atrium, the anterior tricuspid leaflet was incised along its base. A Teflon patch was inserted in a fashion similar to case 1. The immediate postoperative course was smooth and the patient was recatheterised on the fourteenth day after operation. The right ventricle revealed normal function, and there was some regression of the pulmonary hypertension. Left heart study was not performed but recirculation revealed optimal size of the LV outflow tract (fig 1).

Discussion

From the haemodynamic viewpoint, straightforward double outlet right ventricle poses the same pulmonary hypertensive problems as isolated, large VSD. To the best of our knowledge the only work that demonstrated the detrimental effect of right ventriculotomy on the heart with pulmonary hypertension, was that of Stirling *et al.*¹¹ Despite the fact that in more recent years a number of studies concerning surgery of VSD in the infant have been published,^{7-10 12 13} to date there is no

controlled study that proves the superiority of either ventricular or atrial approach. However, it is of interest to note that of the last six major reports on surgery of VSD in infants,^{7-10 12 13} five prefer the atrial approach.^{7-10 13} The authors, too, routinely repair VSDs in infants through the right atrium and recently this technique was also applied to three selected infant cases of DORV.

In order to analyse the anatomical conditions that allow a transatrial approach to DORV, a short review of the current concepts of DORV is necessary. There are a number of classifications of DORV,¹⁴⁻¹⁸ but the relevant point of this report is the relationship of the great arteries.

There are two fundamental types of DORV. The first is DORV with normally related great arteries (NGA)^{14 18 19} and the second is DORV with malposition of the great arteries.^{14 18}

In DORV with NGA except for the presence of subaortic conus,¹⁴ the aortico-left ventricular relationship is similar to that seen in tetralogy of Fallot. Of the 70 DORV cases reported recently from the Mayo Clinic¹⁸ there were two such cases, while in our surgical experience at the Sheba Medical Centre, three of the DORV had NGA. In all the reported^{14 18} and the present cases, the VSD in this particular type of DORV was large and closely related to the aorta (perimembranous VSD).

As one can judge from the postoperative angiograms of the present cases of DORV with NGA (cases 1 and 2), there was an optimal width of the tunnel. It should be mentioned here that in tetralogy of Fallot, where the fundamental relationship between the aorta and the VSD is similar to DORV with NGA, a convenient transatrial repair has been reported.²⁰

The problem is more complicated in DORV when the great arteries are not normally related, as in case 3. The postoperative LV angiogram of case 3 indicates that in the classical form of DORV with side-by-side great arteries, a good sized tunnel can be constructed via the right atrium. But for this, the VSD should be subaortic and the crista should be hypoplastic. It is assumed that the larger the crista and/or the more the aorta is anteriorly placed, the less is the chance of obtaining an adequate tunnel by the atrial approach (fig 2).

Another crucial factor, which was not present in this series, is the infundibular stenosis. Edmunds *et al.*²⁰ repair the infundibular stenosis via the right atrium, but we do not have that experience.

Summing up the above data, there are four factors that must be taken into consideration for the transatrial approach in DORV: the relationship of the great arteries,^{14 18} the location of the VSD,⁸ the size of the crista supraventricularis, and the presence

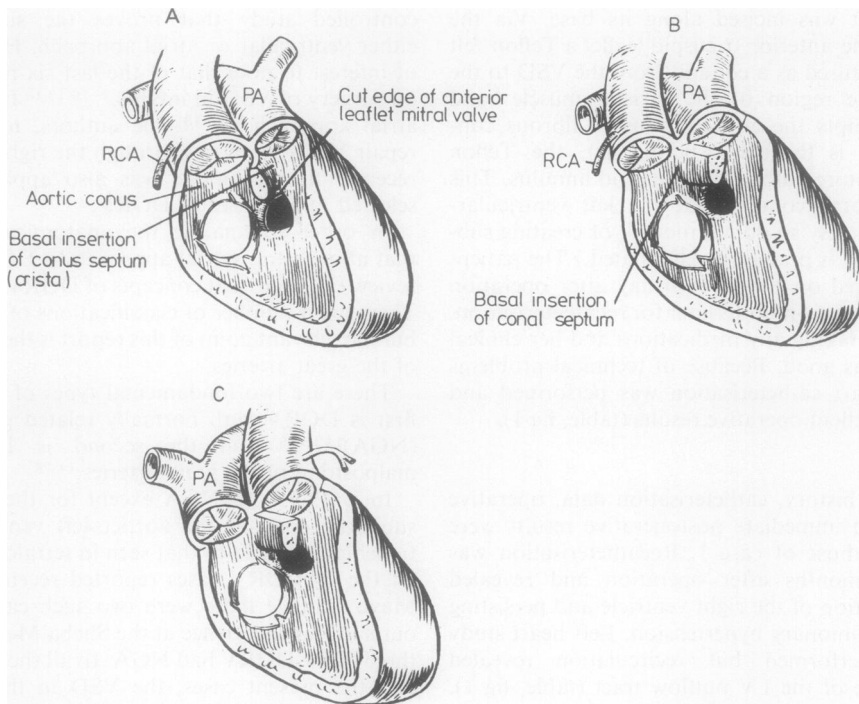


Fig 2 Diagrammatic illustration of the three fundamental positions of the great arteries and crista supraventricularis (conal septum) in DORV. (A) DORV with NGA. (B) DORV with side-by-side location (d-malposition) of great arteries. Notice the fundamental difference in the orientation of the crista (conal septum) between A and B. In A, the septal insertion of the crista is aligned as in tetralogy of Fallot with the anterior edge of the VSD, while in B it is aligned as in transposition of great arteries with the atrial end of the ventricular septum. Hence, the more the aorta is anteriorly placed—that is, oblique or anterior-posterior relationship—and the longer the aortic conus, the more does the crista obstruct the way between a perimembranous VSD and the aorta. (C) DORV with l-malposition. The cono-ventricular anatomy is the same as in B, except for the reversed relationships of the great arteries.

or absence of infundibular pulmonary stenosis. All these can be determined by angiocardiography. The more complicated the DORV, the greater the indication for a transventricular approach.

It should be recalled that in DORV with malposition of the great arteries, the following radiographic and pathological semilunar valve relationships have been described. The classical side-by-side great arteries,^{16 17} oblique and antero-posterior relationships (d-malposition),^{14 15 18 21–25} and l-malposition.^{26–29}

In addition to the different locations of the VSD in the septum, the relationships between the VSD and the great arteries often depend on the conal anatomy. In other words, a VSD in the same perimembranous location can relate differently to the great arteries, depending on the spatial position and

length of subaortic or subpulmonary conus.

In conclusion optimal transatrial repair of DORV can be carried out in the infant under the following circumstances: (1) DORV with normally related great arteries, perimembranous (subaortic) VSD and no infundibular pulmonic stenosis; (2) DORV with side-by-side (d-malposition), perimembranous (subaortic) VSD, small crista supraventricularis, and no infundibular pulmonary stenosis. All other varieties of DORV should probably be repaired through the ventricle.

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