NEW FRONTIERS IN CARDIO-VASCULAR SURGERY

SURGICAL PROSPECTS IN COMPLEX CYANOTIC CONGENITAL HEART DISEASE

By Brian Barratt-Boyes

In view of the many advances in the treatment of complex cyanotic congenital heart disease, we now undertake full investigation of all cyanotic infants regardless of their apparent complexity or age, in the hope that with the detailed analysis of the anatomy that is now possible using modern angiocardiographic techniques, particularly cineangiography in the infant group, a primary corrective procedure can be planned and successfully executed. In this presentation I will emphasise the possibilities provided for primary repair in infancy, using a technique of profound hypothermia produced by surface cooling, followed by circulatory arrest and rapid rewarming with extracorporeal circulation. The possibility of primary repair, regardless of age or complexity of lesion, and the avoidance therefore of a two-stage approach, is a fascinating prospect which deserves our close attention.

Let us look first briefly at a classification of cyanotic congenital heart disease.[1] For cyanosis to occur there are two prerequisites, firstly a communication between pulmonary and systemic circuits and secondly some additional obstruction which causes blood to flow through this defect from right to left.

In the first group (Table I) where cyanosis is determined entirely by obstruction to pulmonary blood flow, we are not concerned with category A, where the obstruction lies in the pulmonary vasculature. In category B, where the obstruction is intracardiac, group 1a includes the large group of infants with tetralogy of Fallot, as well as those with pulmonary atresia and a large ventricular septal defect (VSD) or pseudo-truncus. We will discuss these in more detail later. Where there is pulmonary atresia with intact ventricular septum and shunt reversal at atrial level, treatment remains largely palliative, as it does in tricuspid atresia. We will not discuss Ebstein's anomaly or under-developed right ventricle.

In the second group (Table II) where cyanosis is determined in part by factors other than obstruction to pulmonary blood flow, which may of course be an additional factor in all the conditions listed, we will comment further on truncus, total anomalous pulmonary venous connection, transposition and atrial and ventricular inversions. We should note, however, that in complete A-V canal, the treatment of choice is also complete correction in infancy.

Tetralogy of Fallot and its Variants

In babies born with severe tetralogy and cyanotic spells, routine management has consisted of shunt palliation in infancy followed by repair at five to ten years of age. Analysis of the results achieved with this form of management[2,3] indicates that the ultimate survival of severely affected infants is, at best, about 80 per cent. There is a 10 per cent to 15 per cent mortality or more from shunt surgery and a 5 per cent to 8 per cent mortality at second-stage repair. There is, in addition, a small mortality during the waiting period and a significant morbidity from shunt failure, stroke, cerebral abscess, residual cyanosis and recurrent respiratory infections.

Because of these imperfect results we have, over the last three years, performed a primary intracardiac repair in all tetralogy infants who would otherwise require shunting procedures, using the techniques of profound hypothermia. No infant has been refused repair because of unfavourable anatomy.

The results achieved are shown in Table III. The 5 per cent hospital mortality is clearly superior to that achieved with two-stage management and comparable to that which we obtain with complete repair in older patients. In the 19 survivors there have been no residual shunts or complete heart blocks. One has unfortunately died late of epidemic diarrhoea; a death unrelated to the heart which was well repaired. It is our intention therefore to continue this policy in the expectation that the results will be greatly superior to two-state surgical management.

A similar policy is also followed in infants with double outlet right ventricle and pulmonary stenosis who demand treatment. Two of three with this condition have survived early repair.

There are three other tetralogy variants which require mention:

(a) **Tetralogy of Fallot with absent left pulmonary artery (Fig. 1)**

The right lung in this condition seems to develop an increased vascular resistance inordinately early in life and a shunt to the right pulmonary artery is difficult and risky to perform and gives poor palliation. Moreover, second-stage repair is usually unsuccessful. Early one-stage repair would therefore seem preferable in this group, although to date we have not had the opportunity to test this hypothesis.

(b) **Tetralogy of Fallot with absent pulmonary valve (Fig. 2)**

These infants may develop congestive heart failure early in life and palliation is not possible. Intracardiac repair with VSD closure and outflow tract reconstruction with a valved aortic homograft is the only feasible treatment. The 14-month-old infant whose heart is depicted in Fig. 3, unfortunately died postoperatively of a fulminating tracheobronchitis which was present but unrecognised preoperatively.

(c) **Pulmonary atresia with large VSD (extreme tetralogy or pseudo-truncus)**

This anomaly can be subdivided according to the source of blood supply to the pulmonary arteries from the aorta and to whether or not the left and right pulmonary arteries are confluent. In most of these infants presenting early in life, the pulmonary arteries are confluent and are supplied by a patent ductus (Fig. 4). Unfortunately, duct closure usually occurs with fatal results and treatment is therefore an urgent matter. Palliative shunt surgery is again the usual management, but the mortality is considerably higher than in tetralogy. For this reason we have also attempted one-stage intracardiac repair in such infants, using a valved aortic homograft to reconstitute the pulmonary outflow, combined with VSD and ductus closure.

The results achieved so far have been encouraging (Table IV). Two of the six infants had been palliated in early life (when profound hypothermia techniques were not available) but required repair at 15 and 17 months because of progressive, severe cyanosis. In the first infant to have this repair, a pericardial tube was used. This has subsequently dilated and although he remains well, it is clear that a valve should always be inserted. Note that the diameter of the aortic homograft has been surprisingly large and should allow for considerable, if not complete growth.

Turning now to those conditions where cyanosis is determined primarily by free intracardiac mixing.

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TABLE I

CYANOSIS DETERMINED ENTIRELY BY SEVERITY OF OBSTRUCTION TO PULMONARY BLOOD FLOW

A. **EXTRACARDIAC obstruction**
(R→L at great vessel, ventricular or atrial level)
1. Elevation of PVR (Eisenmenger)
2. Peripheral PA stenoses

B. **INTRACARDIAC obstruction**
1. **Valve or outflow tract stenosis or atresia**
(a) Pulmonary (R→L at ventricular level
R→L at atrial level)
(b) Tricuspid (R→L at atrial level)
2. **Other causes of "obstruction" (R→L at atrial level)**
(a) Ebstein's anomaly of tricuspid valve
(b) Underdevelopment of right ventricle

TABLE II

CYANOSIS DETERMINED IN PART BY FACTORS OTHER THAN SEVERITY OF OBSTRUCTION TO PULMONARY BLOOD FLOW

A. **FREE INTRACARDIAC MIXING**
Truncus arteriosus
Single ventricle
Complete A - V canal
Mitral or aortic atresia
Total anomalous pulmonary venous return

B. **ANATOMICAL DIRECTION OF VENOUS BLOOD TO SYSTEMIC CIRCULATION**
Transposition of great arteries
Atrial and ventricular inversions
Systemic venous drainage to left atrium
Communication between RPA and LA

TABLE III

TETRALOGY OF FALLOT
PRIMARY INTRACARDIAC REPAIR WITH PROFOUND HYPOTHERMIA
JULY 1969 - SEPT. 1972

Age (months)	No.	Hospital Deaths
Under 1	1	0
1 to 2	2	1
3	3	0
4	1	0
6 to 12	8	0
13 to 22	5	0
TOTAL	20	1

TABLE IV

PULMONARY ATRESIA + VSD
PRIMARY INTRACARDIAC REPAIR WITH PROFOUND HYPOTHERMIA

Age (months)	Other Defects	Repair	Outcome
1	PDA	Homograft (17 mm)	Alive
1	PDA	Homograft (19 mm)	DIED (bleeding)
4	PDA	Pericardial tube	Alive
14	PDA	Homograft (16 mm)	Alive
15	Waterston (2 wk) Underdeveloped RV	Homograft (18 mm)	Alive
17	Waterston (3 days) PDA	Homograft (14 mm)	Alive

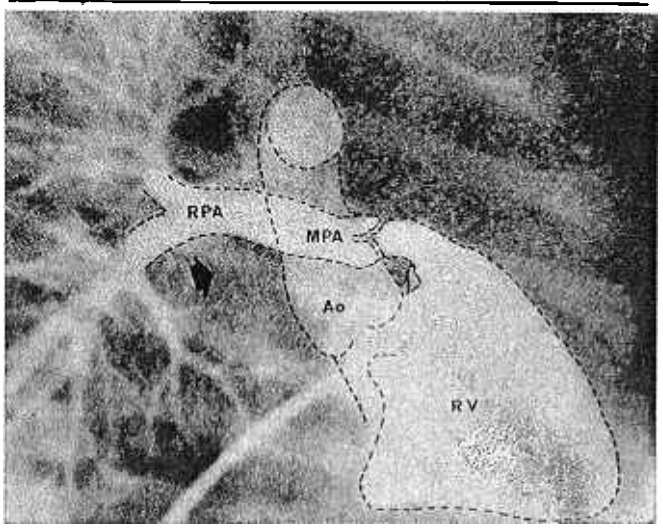

Fig. 1. Cineangiogram frame in right anterior oblique projection demonstrating absence of the left pulmonary artery (the arrow marks the site where this vessel should be visible) in a patient with tetralogy of Fallot. RPA = right pulmonary artery; MPA = main pulmonary artery; Ao = aorta; RV = right ventricle.

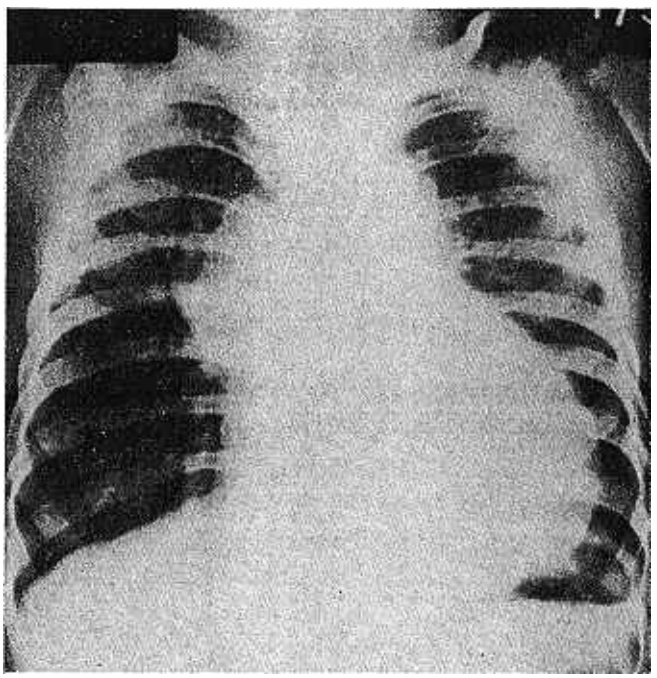

Fig. 2. Chest radiograph of a 14-month-old child with tetralogy of Fallot and absent pulmonary valve demonstrating the aneurysmal dilatation of proximal right and left pulmonary arteries which is a feature of this condition.

TRUNCUS ARTERIOSUS

The intracardiac anatomy in this condition is identical to that in tetralogy with large VSD or pseudo-truncus.[4] The difference lies in the fact that the pulmonary arteries arise directly from the ascending aorta (Fig. 5) and if this orifice is not stenosed there is a high pulmonary blood flow and a sufficient admixture of pulmonary blood to prevent clinical cyanosis. This is a highly lethal condition in infancy due to severe heart failure, sometimes combined with truncal valve incompetence. Banding of the pulmonary arteries is not satisfactory treatment and there can be no argument that early complete repair, if this is possible, would be preferable. The technique of repair is similar to that used in pseudo-truncus.

To date we have operated on four of these infants (Table V). In the first patient, pericardium was used and the absence of a valve was the chief cause of death two months later. The second death was due to damage to the anterior descending coronary artery —an avoidable error. Note that it may be necessary to enlarge the VSD to allow free communication between the left ventricle and truncus artery beneath the teflon patch.

TOTAL ANOMALOUS PULMONARY VENOUS CONNECTION

I mention this condition only to emphasise that with modern surgical techniques the salvage of infants is good regardless of age or site of anomalous drainage. Thus, in those with supracardiac and intracardiac drainage (Table VI), six of seven are alive; and the one death was due to an avoidable error in postoperative management. While in those with infradiaphragmatic drainage, three of four are alive (Table VII).

The management of this anomaly should now consist of atrial balloon septostomy at the time of initial catheterisation followed soon after by intracardiac repair.

Finally, those cyanotic conditions due to anatomical direction of venous blood to the systemic circuit.

TRANSPOSITION WITH LARGE VSD

In simple transposition, immediate balloon septostomy is now followed by atrial baffle repair at three months of age, using profound hypothermia, and with this form of management the salvage of these infants now lies between 85 per cent and 90 per cent.[5]

When a large VSD is present, standard treatment has been to band the pulmonary artery to control heart failure and prevent the development of pulmonary vascular disease, delaying intracardiac repair to an older age. With this two-stage management in our clinic, salvage has been only 12 per cent.[5] Accordingly, over the last three years we have also brought these infants forward for early repair, namely, VSD closure and insertion of an atrial baffle.

As can be seen from Table VIII, the results have been disappointing, particularly in the first month of life. The single hospital survivor from the neonatal group has recently died, nine months postoperatively for reasons which are not clear, as the repair looked intact at necropsy. The results beyond the neonatal period have been more encouraging, although again two of the four hospital survivors have recently died late, possibly from arrhythmia. Currently, we are attempting to hold these babies with intensive medical treatment until they are three to six months old, when repair will be undertaken. Pulmonary artery banding may occasionally be necessary in the neonate, when congestive heart failure cannot be controlled medically.

VENTRICULAR AND ATRIAL INVERSIONS

This can be a most confusing subject and I hope to simplify it by presenting several examples in diagrammatic form. Good quality angiocardiography is essential in order to define the position of each of the four heart chambers. It is essential to know whether there are two ventricles of approximately equal size and not a single ventricle, and to establish that each ventricle has its own atrioventricular valve. We must also know the type of pulmonary stenosis and the position of any VSD. Provided there are two ventricles and two A-V valves, repair should be possible using either an external valved conduit or an atrial baffle.

Fig. 6 establishes the pattern present in the usual variety of D loop transposition with the morphological right ventricle on the right. The atria are concordant with the ventricles and they are also concordant with the viscera, the right atrium and the great veins lying on the same side as the liver. The only anomaly is transposition of the great arteries which is best defined as origin of the aorta from the morphological right ventricle and of the pulmonary artery from the left ventricle. In this diagram the pulmonary artery partly arises from the right ventricle and by definition therefore this becomes a malposition of the great arteries rather than a transposition. These anomalies with malposition are usually called double outlet right or left ventricle and this particular example is a variety of double outlet right ventricle. It is best treated by closing the VSD so that the pulmonary artery arises entirely from the left ventricle and then inserting an atrial baffle.

Fig. 7 shows two examples of so-called corrected transposition. In the first example, the atria and viscera are normally placed and therefore concordant, while the ventricles are reversed or inverted and therefore discordant with the atria. Because there is also transposition, the circulatory pathways remain normal. Cyanosis will be present in this condition when there is a VSD and pulmonary stenosis. As the pulmonary artery arises posteriorly from the morphological left ventricle and the stenosis often involves the pulmonary valve and subvalve area, it may not be possible to relieve it directly. Treatment then consists of a valved conduit between the left ventricle and pulmonary artery and closure of the VSD. The second example is a mirror image of the first. Here there is inversion of the atria and the viscera, so that while they remain concordant, the right atrium lies on the left. The ventricles are discordant with the atria and there is transposition. As there is also pulmonary stenosis and a VSD, there is cyanosis. Treatment is identical to the last patient, except that the conduit lies on the left of the body rather than the right. This seven-year-old boy has made a good recovery.

The final diagram shows a most unusual heart which breaks all Dr. Van Praagh's rules. Thus, there is visceroatrial discordance, a most unusual finding, with the right atrium on the left but the liver in normal position on the right. The ventricles are discordant with the atria but there is no transposition. Physiologically this heart is similar to D loop transposition and treatment consisted therefore, in this two-year-old child, of performing an atrial baffle repair through the leftsided right atrium. She made a good recovery.

I hope that this type of approach will help in analysing these problems and deciding whether treatment is feasible and what it should be. Dextrocardia may also be present, as it was in this last child, but it is really of no importance.

SUMMARY

The possibilities of correction of cyanotic congenital heart disease in the infant period are presented. Emphasis is placed on the management of tetralogy of Fallot and its variants with absent left pulmonary artery, absent pulmonary valve, and pulmonary atresia with confluent pulmonary arteries. Truncus arteriosus, total anomalous pulmonary venous connection and complex transposition are also discussed. Finally, a number of examples of ventricular and atrial inversion with cyanosis are presented in diagrammatic form.

TABLE V

TRUNCUS ARTERIOSUS
PRIMARY INTRACARDIAC REPAIR WITH PROFOUND HYPOTHERMIA

Age (months)	Aortic Incompetence	Repair	Outcome
1	Mild	Homograft (14 mm)	Alive
2	Mild	Homograft (19 mm) VSD enlarged	Alive
2	Absent	Pericardial tube	Dilatation pericardium Reop. age 4 months DIED
3	Moderate	Homograft (18 mm) VSD enlarged	DIED (technical error)

TABLE VI

TOTAL ANOMALOUS PULMONARY VENOUS CONNECTION
REPAIR WITH PROFOUND HYPOTHERMIA

Age	Type of Drainage	Outcome
8 days	Coronary Sinus (+ interrupted arch + VSD)	Alive
2 months	Right SVC	Alive
3 months	Right SVC	DIED (pneumothorax)
3 months	Coronary Sinus	Alive
4 months	Left SVC	Alive
8 months	Mixed	Alive
10 months	Coronary Sinus	Alive

TABLE VII

INFRADIAPHRAGMATIC TAPVC
REPAIR WITH PROFOUND HYPOTHERMIA

Age	WT	Outcome
5 days	2·7 kg	Alive
6 days	2·6 kg	Alive
8 days	3·3 kg	DIED at 4 months (sub-glottic stenosis)
3 months	4·4 kg	Alive

TABLE VIII

TRANSPOSITION WITH LARGE VSD

Age	VSD	Outcome
10 days	Type 2	DIED 12 hrs/low output
12 days	Type 3	DIED 12 hrs/ 'tamponade'
19 days	'A - V Canal'	DIED 2 days/ surgical error
27 days	Taussig Bing	DIED 24 hrs/low output
27 days	Type 2 + PDA	Alive
29 days	Type 2 + PDA	DIED Theatre/ surgical error
2 months	Type 2	Alive
2 months	Type 2	DIED 6 days/cardiac arrest
2 months	Type 2	Alive
3 months	Type 2	Alive
4 months	'A - V Canal'	Alive
6 months	Type 2 + PDA	DIED 24 hrs/bleeding

Fig. 3. Autopsy specimen of the child whose chest radiograph is shown in Fig. 2. The distal right ventricle and proximal pulmonary arteries have been opened anteriorly to display the aortic homograft and the enormously dilated proximal pulmonary arteries.

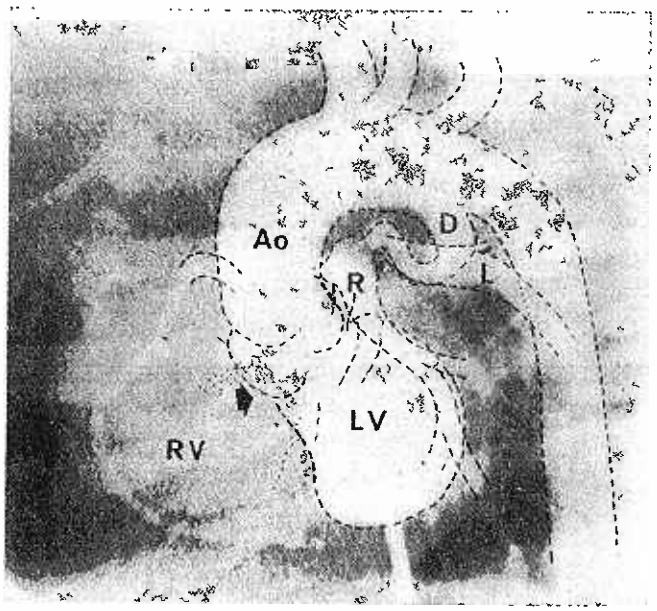

Fig. 4. Cineangiogram frame in left anterior oblique demonstrating confluent left and right pulmonary arteries in a patient with atresia of the pulmonary valve and pulmonary trunk and large ventricular septal defect (arrow).

Ao = aorta; D = patent ductus arteriosus; R = right pulmonary artery; L = left pulmonary artery; LV = left ventricle; RV = right ventricle.

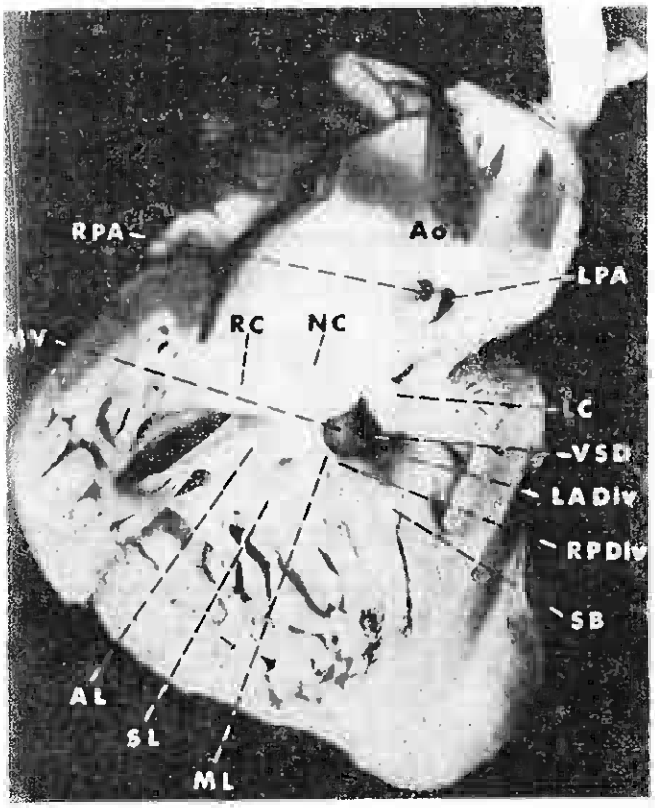

Fig. 5. Autopsy specimen of truncus arteriosus to show the anatomy of the ventricular septal defect (VSD) and the origin of left and right pulmonary arteries (LPA, RPA) from the left postero-lateral aspect of the truncal artery (AoA). From Van Praagh, R. and Van Praagh, S. Amer. J. Cardiol., 16, 406, 1965. Reproduced with permission.

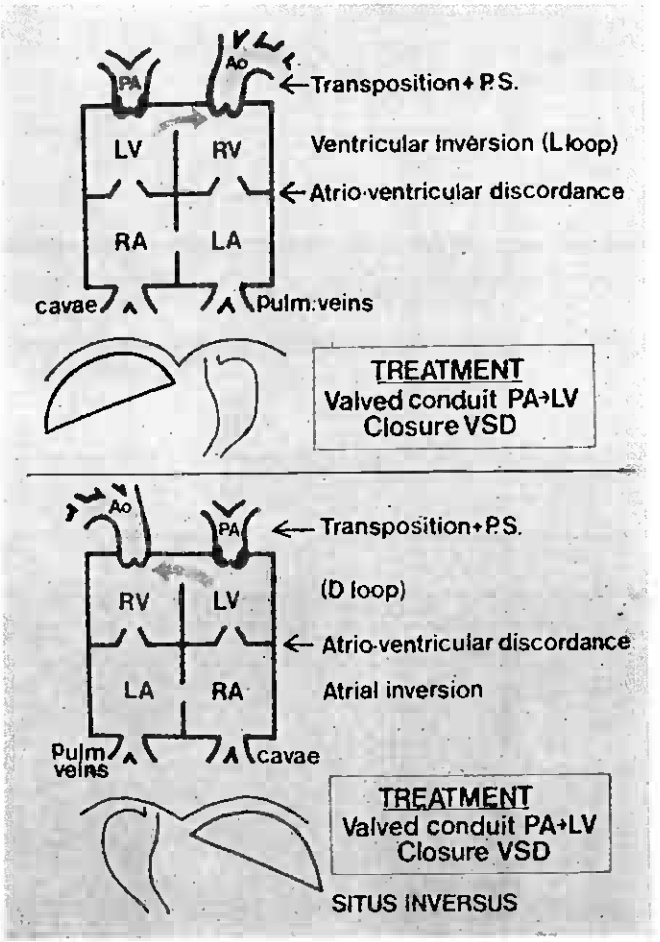

Fig. 7. Diagrammatic representation of two varieties of physiologically corrected transposition with pulmonary stenosis (PS) and ventricular septal defect.

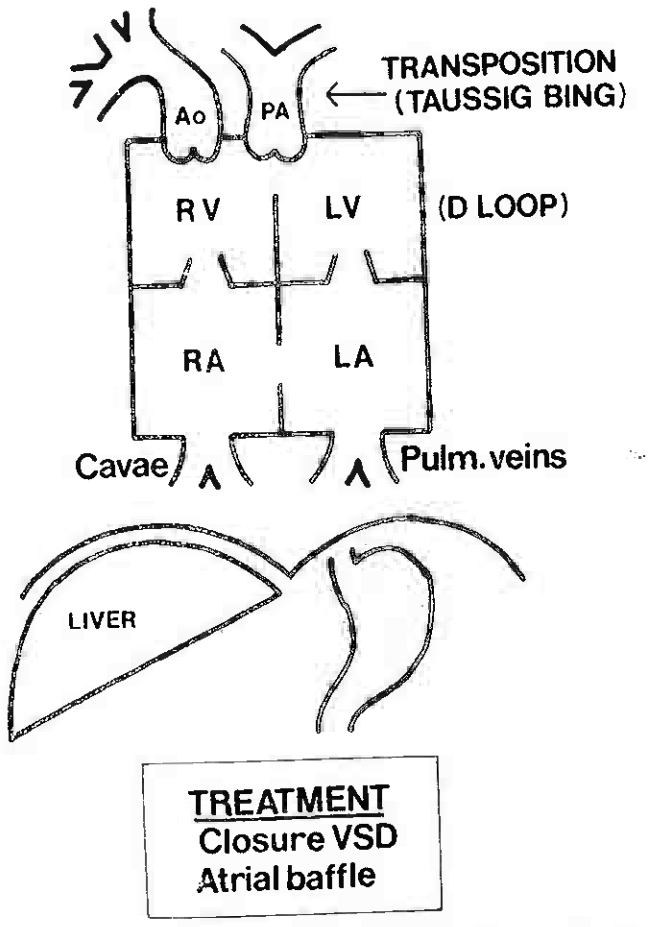

Fig. 6. Diagrammatic representation of Taussig-Bing anomaly. (for abbreviations see other figures).

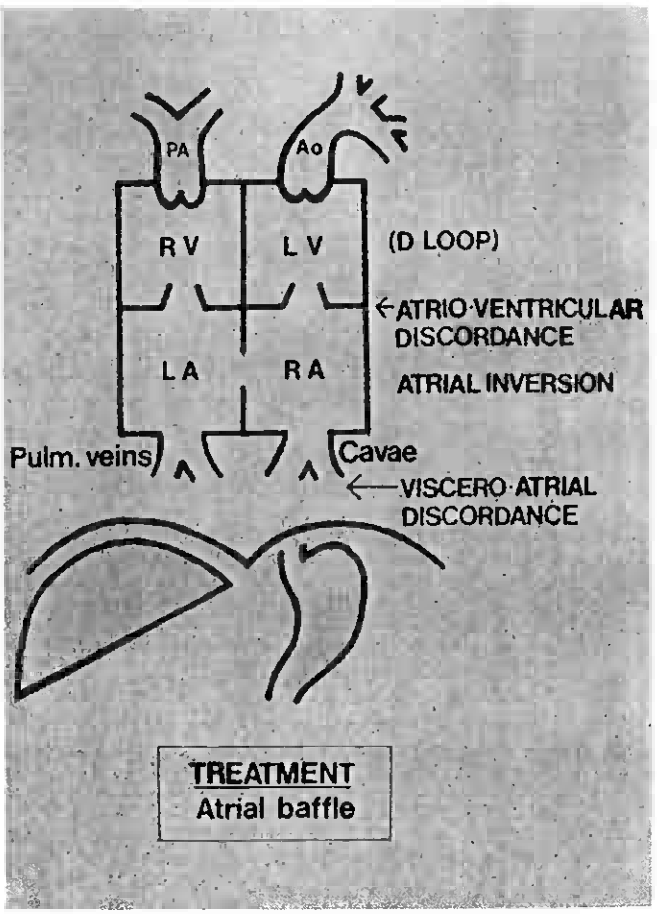

Fig. 8. Diagrammatic representation of isolated atrial inversion.

It is clear from this analysis that surgical treatment of complex cyanotic heart disease has improved and that with the advent of techniques which allow one-stage repair in infancy regardless of the complexity of the lesion, future management of many of these conditions will be simplified for both the patient and the parents if not for the surgeon.

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