TRANSPOSITION OF GREAT ARTERIES : A PRELIMINARY REPORT ON LOCAL EXPERIENCE WITH BALLOON ATRIAL SEPTOSTOMY

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SUMMARY

Eleven consecutive cases of simple transposition of great arteries were palliated by balloon atrial septostomy. The arterial saturation was improved and the gradient across the atrial septum was reduced or abolished. Improvement in the clinical status was achieved in all the patients. There was no mortality and morbidity was mild. A description of the technique as well as a brief review of the anatomy and haemodynamics of complete transposition of great arteries are the aims of this paper.

INTRODUCTION

Since the first description of Transposition of Great Arteries (TGA) by Baillie in 1797,[1] this complex defect has been the subject of numerous research projects and articles. This is perhaps a reflection of the complexity and uniqueness of its anatomy and physiology. At first glance the defect seems incongruous and is "impossible" for survival. However, against all odds it does occur! An incidence of about 0.2 per 1,000 live births or 10.5% of all congenital heart diseases[2] have been reported. Liebman[3] in a large review of its natural history, estimated an incidence of 1 per 4,500 births. It is in fact the second most frequent of the congenital heart defects seen.[2,4] In view of the systemic and pulmonary circulation being in parallel circuits rather than in series as is normal, survival without intervention is bleak. 89.3% are dead by one year of age and life expectancy at birth is only 0.65 years.[3]

Survival was remarkably improved by the introduction of the ingenious surgical techniques[5-8] and most recently Jatene.[9] However the most significant development in the salvage of these infants is the equally ingenious technique of the Balloon Atrial Septostomy (BAS) introduced by Rashkind.[10]

This paper describes the initial experience with this procedure to palliate infants with TGA and its subsequent introductions as a routine service in the Kuala Lumpur General Hospital.

MATERIALS AND METHODS

We will first clarify our definition of TGA as it has been defined differently in the past by various workers. This problem had been highlighted by Shaher[11] in an editorial and also more recently by Tynan.[12] These definitions were so varied that confusion arose as to what is TGA? It was somewhat refined by Van Praagh[13] in 1971. Using the sequential segmental analysis[14,15] and the recommendation of Tynan,[12] complete TGA is defined as those hearts with atrio-ventricular concordance and ventriculo-arterial discordance, i.e. morphological right atrium connecting to the morphological right ventricle and the aorta arising from the morphological right ventricle. The morphological left atrium connects to the morphological left ventricle and the pulmonary artery arising from the morphological left ventricle.

Cases of TGA with intact ventricular septum, small significant ventricular septal defect or closing

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ductus arteriosus were classified as simple TGA. Those cases with significant additional defects such as large ventricular septal defect, pulmonary stenosis, aortic stenosis or coarctation of aorta were classified as complex TGA.

This report is based on patients with simple TGA. The case notes and cardiac catheter reports of the first consecutive eleven cases of simple TGA who underwent BAS are reviewed. These cases represent the small group of TGA cases seen at the Department of Cardiology at General Hospital, Kuala Lumpur, which were suitable for palliation by BAS. In each case, the diagnosis was made by 2D echocardiography using a 5MHz transducer (ALOKA SSD-118). The echocardiographic features are as described previously.[16,17] The basic data and clinical features are listed in Table I.

All the patients were subjected to cardiac catheterisation, either on an emergency or semi-emergency basis. Full general anaesthesia were administered in all the cases. Precautions were taken to prevent hypothermia, hypoglycaemia and acidosis. Percutaneous technique was used in case 1 whilst cut down for access to the femoral vein was used for the remaining ten. The protocol for the procedure is as described by Mullins and colleagues[18] with minor modifications to suit local conditions. The diagnoses were confirmed by selective ventriculography. BAS was performed

TABLE I
CLINICAL CHARACTERISTICS OF PATIENTS WITH TRANSPOSITION OF GREAT VESSELS

Age (days)	Sex	Weight (kg)	Presenting problems	Murmur	ECG	CXR	Refering diagnosis	Echo diagnosis	Final diagnosis
120	F	4.7	Cyanosis	Nil	RAD RVH	Cardiomegaly narrow base plethora	TGA VSD	TGA	TGA
6	M	—	Cyanosis	Nil	N	N	CCHD,	TGA DA	TGA DA
49	F	3.0	Cyanosis	ESM	RAD RVH	Cardiomegaly narrow base plethora	P. At.	TGA	TGA
13	F	3.1	Cardiac failure	Cont. M	RAD RVH	Cardiomegaly plethora	PDA	TGA DA	TGA DA
60	M	3.8	Cyanosis cardiac failure	ESM	RAD RVH	Cardiomegaly narrow base plethora	TGA	TGA	TGA
3	M	3.3	Cyanosis	Nil	N	N	TGA	TGA	TGA
31	F	3.0	Cyanosis	ESM	RAD	Narrow base plethora	TGA	TGA	TGA
30	M	3.2	Cyanosis cardiac failure	ESM	N	Cardiomegaly narrow base plethora	TGA VSD	TGA	TGA VSD
4	M	2.8	Cyanosis cardiac failure	ESM	RAD RVH	Cardiomegaly narrow base plethora	TGA	TGA DA	TGA DA VSD
14	F	2.5	Differing cyanosis CCF	ESM	RAD RVH	Cardiomegaly narrow base plethora	TGA CoA DA	TGA AoA DA	TGA DA
42	M	2.7	Cyanosis	ESM	RAD RVH	Cardiomegaly narrow base plethora	TGA	TGA	TGA

ESM = Ejection Systolic Murmur; RAD = Right Axis Deviation; RVH = Right Ventricular Hypertrophy; N = Normal; CCHD = Cyanotic Congenital Heart Disease; P. At. = Pulmonary Atresia; DA = Ductus Arteriosus; CoA = Coarctation of Aorta.

using the Miller Atrioseptostomy Catheter (Miller Balloon Atrioseptostomy Catheter American Edwards Laboratory 83-051-5F) filled with 3 to 4cc of dilute contrast media. On completion of BAS, repeat assessment of left atrium to right atrium pressure gradient and arterial saturation were made to assess the adequacy of the procedure.

All the patients except for cases 1 and 2 were extubated and sent back to their respective wards for observation. Cases 1 and 2 were observed in the intensive care unit.

RESULTS

The cardiac catheterisation data are listed in Table II.

Cases 1 and 2 represent our initial experience and the long duration of the procedure is due to our own learning curve. The mean duration of the procedure is 72.4 min (range 54 to 104 min). The mean duration of the screening time were 7.4 min (range 4.5 to 10 min).

The basic anatomy demonstrated by angiography was as predicted by echocardiography. Ductus arteriosus was correctly predicted in cases 2,4,9 and 10. The small ventricular septal defect in cases 8 and 9 was not detected by echocardiography. In case 11 the presence of coarctation of aorta was wrongly predicted by echocardiography.

BAS was successfully carried out in all using the largest balloon possible, i.e. 3.5 to 4cc (balloon diameter of about 20 to 20.5mm). In cases 9 and 10, the procedure was only successful with a 3cc balloon, not surprising as these two cases were the smaller infants. The results of BAS were assessed by the rise in systemic arterial oxygen saturation, mean pressure gradient across the atrial septum and clinical improvement as suggested by Neches and colleagues[19] (see Table III). The arterial saturations pre and post BAS are shown in Fig. 1. There was a rise in the arterial saturation in all the patients except for case 4 where the pre BAS arterial saturation was already 98.2%. The mean pressure gradient across the atrial septum pre and post BAS is shown in Fig. 2. Using the criteria suggested by Neches and colleagues,[19] all were considered "good results" except for cases 2 and 4 which could only be considered as "fair". In case 2 the arterial saturation increase was only

TABLE II

CARDIAC CATHETERISATION DATA OF PATIENTS WITH TRANSPOSITION OF GREAT ARTERIES

Duration (mins)	Screening time (min)	Pressures (mm Hg) RA	RV	Ao	LA	LV	PA	Saturations (%) SVC	IVC	RA	RV	Ao	LA	LV	Compli-cations	Size of Balloon (cc)
–	–	12	92/8	100/64	20	–	–	46.5	43.5	44.4	56.5	51.6	100.9	–	Nil	4
104	–	8	52/10	52/24	11	–	–	49.6	69.6	70.3	24.9	21.9	102.9	–	CAVB RFV Tear	4
70	7	18	80/0	–	20	50/0	–	57.9	61.6	68.5	67.0	–	102.4	103.3	Nil	4
54	8	4	80/0	80/40	9	50/0	–	70.2	72.5	75.7	98.2	98.2	106.2	106.3	Nil	4
63	4.5	11	82/0	64/52	17	65/0	–	29.5	28.6	31.4	33.6	31.9	98.3	96.3	Nil	4
64	8	12	54/0	–	12	36/0	–	–	–	–	–	29.4	–	–	Hypotension Hypoglycae-mia Idionodal rhythm CAVB	4
73	10	8	70/5	68/58	8	84/5	–	34.4	55.9	41.6	37.5	38.5	101.6	101.0	Nil	3.5
67	7	11	70/4	70/40	13	60/4	–	47.2	48.2	60.4	65.7	66.9	–	103.7	Nil	3.5
65	10	6	80/0	80/50	11	80/0	–	56.6	57.7	54.1	64.5	65.2	104.0	107.7	2:1 AVB	3
99	6	11	58/4	55/22	13	56/6	–	45.5	78.0	61.5	60.7	53.9	99.6	100.7	2:1 AVB RFV Tear	3
65	6	20	60/0	60/36	20	40/2	–	36.6	34.4	43.4	45.7	46.9	95.5	99:3	CAVB	4

CAVB = Complete AV Block; RFV = Right Femoral Vein; AVB = AV Block.

TABLE III

DATA ON PATIENTS WITH TRANSPOSITION OF ARTERIES

Oxygen Saturation rise (%)	Post BAS LA/RA Gradient (mmHg)	Results	Clinical improvement
51.4	–	Good	Yes
7.4	–2	Fair	Yes
44.8	2	Good	Yes
–	3	Fair	Yes
77.0	0	Good	Yes
139.5	–1	Good	Yes
91.7	1	Good	Yes
29.6	1	Good	Yes
39.4	0	Good	Yes
31.0	0	Good	Yes
70.4	0	Good	Yes

7.4% whilst in case 4, the mean atrial gradient remained at 3 mmHg post BAS. All patients irrespective of the post BAS saturation and atrial gradient improved clinically. This was particularly so in the improvement in the colour and decrease in tachypnoea.

Repeat echocardiography confirmed a tear in the septum primum in all the cases. The echocardiographic picture of the atrial septum pre and post BAS in case 3 is shown in Fig. 3.

DISCUSSION

Transposition of great arteries (TGA) is the second most frequent congenital heart disease and the most frequent of the cyanotic congenital heart disease.[2,4] However this has not been our experience. This is an apparent observation as we believe that most of the cases had succumbed before being referred to us. We hope this short paper would alert our paediatric colleagues to the problems of TGA and that there are now avenues to save them.

The foetal physiology of TGA is obviously compatible with survival of the foetus as the patients were not stillborn.[20] However, after birth, the separation of the pulmonary and systemic circuit would result in two parallel circulations in TGA in contrast to the series circulation in the normal. Severe hypoxia would occur unless there are anatomic communications to allow intercirculatory mixing. Mair and colleagues[21] had highlighted that the number, size and position of the communications are important factors in determining mixing and hence final arterial saturation in the neonate. They also stress the importance of the creation of an atrial septal defect for the survival of those infants with TGA and poor mixing. In view of the parallel circulation, any shunting across the communications must be bi-directional and balanced over a time frame. Otherwise, one circuit would empty into the other. It is this shunting that is important in determining survival and they are termed the effective pulmonary blood flow (QEP) or effective systemic blood flow (QES) (Fig. 4).

The important point in BAS is to create an atrial septal defect by tearing the septum primum and not just mere stretching of the fossa ovalis. Thus the balloon should be withdrawn across the atrial septum in a single rapid jerk. The initial recommendations were to use a balloon filled with 2cc of dilute contrast. With improvement in design and materials, it is now possible to use a balloon with 4cc capacity. We chose the newer balloons in order to achieve a bigger tear of the septum primum.

We do not routinely make any effort to enter the pulmonary artery during the initial cardiac catheterisation as it was felt that it was not necessary at that moment. The prime concern was rapid diagnosis and early BAS before clinical

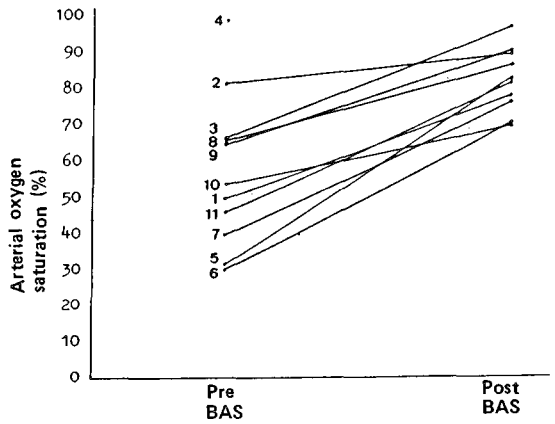

Fig. 1

Arterial oxygen saturation before and after balloon atrial septostomy (BAS) (repeat saturation studies not done).

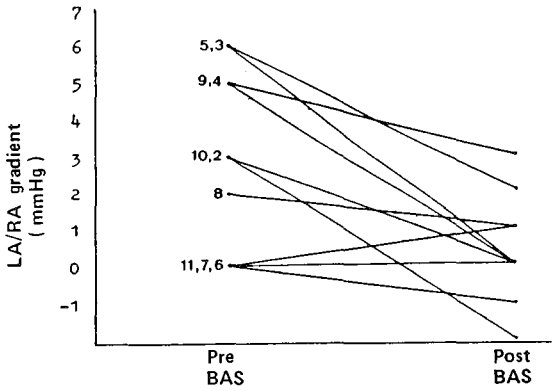

Fig. 2

LA/RA Gradient before and after balloon atrial septostomy (BAS). (NB: Repeat pressure studies were not done in case 1.)

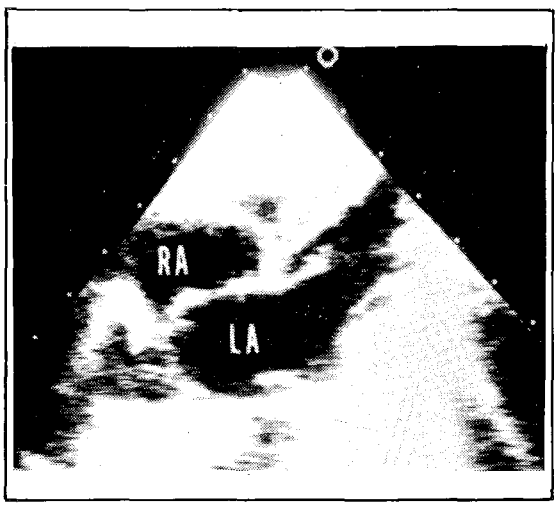

Fig. 3A Pre BAS: Atrial septum intact.

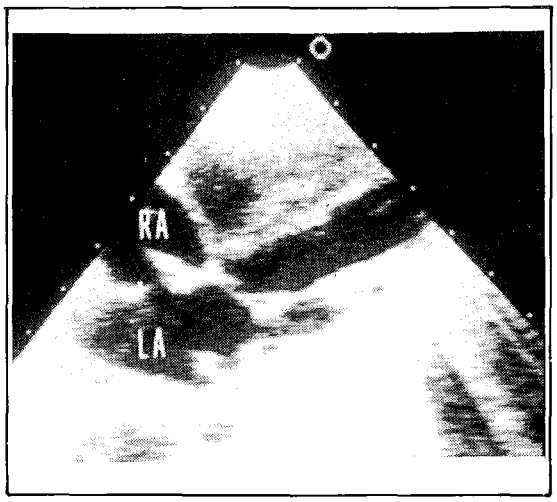

Fig. 3B Post BAS:ASD created: cross marks position of ASD.

deterioration occurred. Moreover, most of the patients would require a repeat cardiac catheterisation study at a later date prior to definitive surgery.

The most frequent complication we encountered was transient heart block which occurred in five of our patients. The femoral veins in two of our patients (cases 2 and 10) tore during the procedure and could not be repaired. In both these cases, the veins were ligated and apart from some transient venous congestion, no short term untoward effects were seen. Perhaps better care in the introduction of the balloon would avoid this problem in the future.

Though our numbers were small, we were encouraged by the results achieved. Particularly so, with the improvement observed during the clinical state. Our main concern was the older age of some of our patients. This had a number of disadvantages i.e. they are often severely cyanosed and hypoxic and there may be muscularisation of the septum primum making BAS difficult. Case 1 is an example who presented at the age of four months! She underwent successful Mustard's operation within two weeks of BAS and the atrial septal defect created was reported by the surgeons as inadequate. Perhaps earlier referrals would obviate this problem.

The introduction of BAS by Rashkind[10] was indeed the most significant development in salvaging infants with TGA. Even in the late 1960s[22] and

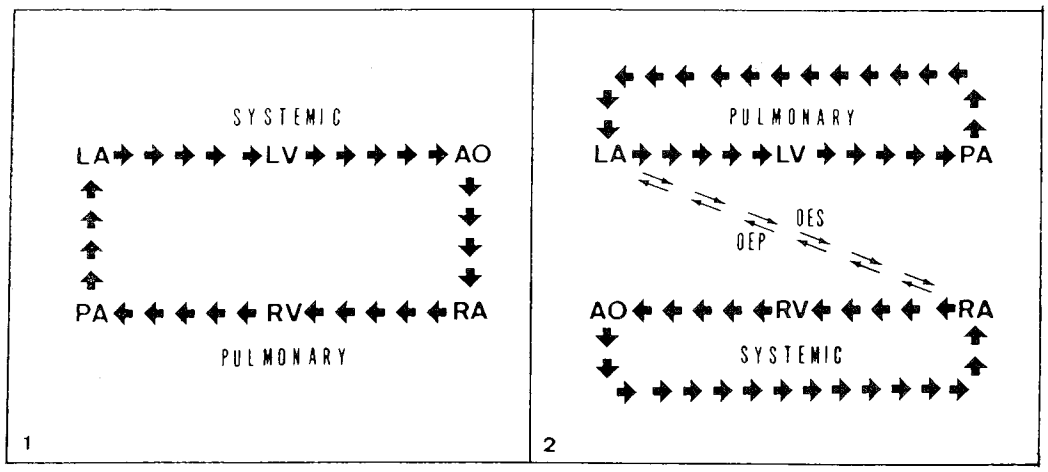

Fig. 4 Haemodynamics in normal and in TGA. 1. Normal, both systemic and pulmonary circulation are in series; 2. TGA, the systemic and pulmonary circulation are in parallel. The effective pulmonary or systemic blood flow were shown as in the presence of an atrial septal defect. QEP = Effective pulmonary blood flow; QES = Effective systemic blood flow.

the early 1970s,[2 3] consistent good results were achieved. However despite this initial success, those infants are still at risk of complications. Parsons and colleagues[2 3] had reported up to 49% mortality by four years if they did not undergo definitive surgery. Most of the causes of morbidity and mortality were related to the complications of polycythaemia and hypoxia.

Thus, the recommended line of management of patients with simple TGA is early palliation by BAS shortly after birth followed by elective Mustard's or Senning's procedure at six to nine months of age, or earlier, if clinical deterioration occurs.

We report here our initial experience with BAS in palliating infants with simple TGA. Though we report only short term results, those patients who still maintain follow up have all been well. Their growth and developmental milestones have been within normal limits. We believe that this technique should now be introduced as a routine service for those patients with simple TGA who present to us. With the success of this technique, we hope to present to our surgeons these patients who are in their best of health for definitive surgery.

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