INTRACARDIAC REPAIR FOR SINGLE OR COMMON VENTRICLE, CREATION OF A STRAIGHT ARTIFICIAL SEPTUM

By Tatsuta Arai* Shigeru Sakakibara† Masahiko Ando† and Atsuyoshi Takao†

Among the few congenital cardiac malformations for which a difinitive corrective procedure has not been achieved, is single or common ventricle.

This cyanotic or admixture type of lesion has a wide clinical spectrum, ranging from a deeply cyanosed to apparently pink color, and from neonatal death to survival up to 56 years, mainly depending upon the associated anomalies of the outflow tracts to the great arteries. Anatomically, it has been classified into four different types, namely single left ventricle, single right ventricle, undivided ventricles and infundibulum without ventricular sinuses, according to the morphology of the uni-ventricle present.

For the past few years we have been thinking about possible ways to construct an artificial ventricular septum in certain types of single or common ventricle. Because the most essential function of the ventricular septum appears to be the partitioning of the two ventricles in addition to contribution to contractility, conduction pathway and so on. This is suggested by the well maintained cardiac output in these uni-ventricular patients, and also by the reported hemodynamic studies on dogs with subtotal prosthetic replacement of the ventricular septum.

The purpose of this paper is to present a successful intracardiac repair with an artificially created straight ventricular septum, on two patients with single left ventricle (Type A III, solitus, Van Praagh's classification), and on one patient with common undivided ventricles (Type C I, Solitus). The indications for this procedure and further possibility will be discussed.

CASE PRESENTATIONS

Case 1. A 7 year 5 month old female was admitted to the Heart Institute Japan with complaints of moderate severe cyanosis and exercise intolerance. Grade 4/6 systolic ejection murmur was heard along the upper left sternal border with an accentuated single second sound. **The chest x-ray** revealed no cardiomegaly (CTR 53%), decreased pulmonary vascularities and a characteristic convex left upper cardiac border suggesting the presence of l-transposition. **The EKG** showed complete heart block with a ventricular rate of 50/min., frontal QRS axis of + 130 and QRS configurations in the chest leads suggestive of inverted ventricles with a predominant right anterior force.

Cardiac catheterization revealed a patent foramen ovale, and moderate pulmonary valvular stenosis with a systolic pressure gradient of 90 mm Hg.

Angiocardiograms at the ventricle, left atrium and aortic root established the diagnoses of single left ventricle with rudimentary outlet chamber, l-transposition of the great arteries (l-TGA'S), and situs solitus of viscera and atria (ie Type A-III Solitus). A single right coronary artery and right aortic arch were also opacified. Moderate pulmonary stenosis was present with poststenotic delatation of the pulmonary artery. During early systole of the ventricular injection two negative shadows were seen in the ventricle, suggesting the presence of two functioning atrio-ventricular valves (A-V valve) (double inlets left ventricle). The great arteries were almost side by side, with an enlarged aorta to the left of a moderately hypoplastic pulmonary artery. The former appeared to be more posterior and inferior compared to that of the usual type of transposition, and to override upon a large bulbo-ventricular foramen (VSD).

*Jikei University School of Medicine.
†Heart Institute Japan, Tokyo women's Medical college.

These findings, ie well divided atrio-ventricular valves, and l-TGA'S with side by side great arteries, were all favorable for the creation of a straight artificial septum which could separate the pulmonary and systemic circulations with the appropriate venous blood. Besides this, the pulmonary vascular bed had been protected from developing vascular obstruction by the moderate pulmonary stenosis.

Cardiac Surgery: The heart was exposed through a median sternotomy. The external view of the heart was as expected from the angiocardiographic studies. Immediately after its origin from the aorta, the single right coronary artery divided into a large right, anterior descending and small left circumflex coronary artery. Under extracorporeal circulation, a longitudinal ventriculotomy measuring 4 cm was made along the avascular area between the anterior descending and the preventricular branch of the right coronary artery.

Inspection through the incision revealed typical left ventricular morphology with a smooth septal surface, and obliquely and finely trabeculated apical portion. The right and left A-V valves were well formed and opened into the large left ventricle (Double inlets left ventricle). The papillary muscles to each A-V valve were separated by a so called posterior median ridge, running obliquely from the medial junction of the A-V valves toward the cardiac apex along the posterior wall of the left ventricle. Beyond the hypoplastic ventricular septum on the left upper portion, a rudimentary outlet chamber with a small trabeculated apical recess was recognized through a large subaortic VSD.

Pulmonary stenosis was valvular due to a bicuspid valve, and subvalvular fibrous probably due to a jet lesion. The hypoplastic pulmonary valve was just above the medial corner of the right A-V valve, and a direct fibrous continuity was present between them. The enlarged aortic valve was upon the rudimentary outlet chamber, but considerably overriden on the ventricular septum. A thin subaortic conus (5 mm in length) separated the aortic valve above and the left A-V valve below. No conal septum was present between the semilunar valves, and they were directly continuous.

An imaginary line connecting the right A-V valve to the pulmonary valve (Sv-PA line=systemic venous to pulmonary artery line) and that connecting the left A-V valve to the aortic valve (PV-AO line=pulmonary venous to aortic line) in this case were parallel. An artificial conal septum supposed between the both semilunar valves and an artificial ventricular sinus septum supposed between both the A-V valves were on the same plane, and just between the above mentioned imaginary lines. Hence creation of a straight ventricular septum was possible in this case.

After release of pulmonary stenosis and enlargement of the VSD by removal of the supero-anterior rim, to secure the size of the aortic outflow and to incorporate the apical recess into newly made pulmonary venous ventricle, a straight ventricular septum was created with an ovoid teflon patch. The most important part is to avoid injury on His bundle and left side bundle branches to the papillary muscles of the left A-V valve because these conducting pathways are for the newly made pulmonary venous or systemic ventricle. To avoid the above-mentioned conduction ways, the artificial sinus septum was laid along the root of the medial leaflet of the right A-V valve with interrupted mattress sutures until it emerged at the posterior leaflet of the same valve which attached to the vestibulum of the coronary sinus where A-V Node is assumed to be present. After passing the coronary sinus vestibulum at the posterior corner of the junction between the right and left A-V valve, stitches were con-

tinued along the posterior wall on the right side of the posterior median ridge, until they passed the root of the papillary muscles of the left A-V valve. An artificial conal septum was attached just between the two semilunar valves. The apical and anterior free wall portion of the artificial septum was so fixed as to equally divide the newly made systemic and pulmonary ventricles, and special care was paid to make the septum tight so as not to become excessive during ventricular systole.

Immediately after the procedure, preoperative heart rate (50/min.) was established. However, temporary pacemaker electrodes were implanted to overcome the post-operative critical period. Congestive heart failure and increased pulmonary secretion were managed well by digitalis, diuretics, tracheostomy and external pacing with a rate of 90-100/min. On 26th post-operative day the external pacemaker stopped working due to an electrode failure. The EKG at that time revealed complete A-V block with a ventricular rate of 60/min., (the same as that before surgery), and the QRS duration which was 0.08" before surgery increased to 0.11", probably due to injury on the right side bundle branch by the sutures along the posterior median ridge. As congestive heart failure appeared to increase with her own rhythm, a permanent pacemaker (Device 270 type) with a fixed rate of 81/min. was implanted. Her general condition was improved. The post-operative angiograms at the systemic and pulmonary ventricle revealed almost equal in size without demonstrable ventricular shunt. Residual pulmonary stenosis was visualized at the valve ring. The pressure in the pulmonary ventricle was 120/3 and was almost equal to that of the systemic ventricle. It is now 1 year 8 months post-operatively and she is enjoying cheerful school life.

Case 2. A 7 year 1 month old male with mild cyanosis had an established diagnosis of single left ventricle with a rudimentary outlet chamber, 1-TGA'S and Situs Solitus of viscera and atria (Type A III Slitus). The pulmonary artery had been banded at 3 yrs 6 month of age, because of markedly increased pulmonary flow with congestive heart failure. **The EKG** revealed, 2 to 1 A-V block with a ventricular rate of 75/min., and findings suggesting ventricular inversion. **The chest x-ray** showed moderate cardiomegaly, and increased pulmonary vascularities. Right atrial and ventricular **angiocardiograms** were performed to elucidate the anatomy of the A-V valve and of the aortic outflow. These confirmed the diagnosis. The right and left A-V valves appeared to be well separated. The enlarged pulmonary artery was posterior and to the right of the mildly hypoplastic aorta. VSD appeared to be not small.

Surgery: On opening the chest through a median stenotomy, massive pericardial adhensions were present due to the previous operation. After careful disection, the anterior ventricular surface was exposed. There was a large preventricular branch arising from the proximal portion of the right coronary artery A, for no incision space was obtained between this artery and the anterior descending coronary artery, a longitudinal incision along the right side of the preventricular branch was made. Several papillary muscles to the anterior leaflet of the Rt A-V valve, and an abnormal muscular ridge in the subsemilunar valve area were traversing the incision field. One of the former which was small and accessory was temporarily divided, and the rest were tracted cranially. The latter was resected, for this structure seemed to be an abnormally large trabeculation between the left and right side of the free wall. Typical left ventricular cavity was now in sight. Both A-V valves were well formed and opened into the LV (Double inlets left ventricle). The pulmonary valve was posterior and directly continued to the medial leaflet of the right A-V valve. The aortic valve was supero-anterior and to the left of the pulmonary valve. It was originating from the rudimentary outlet chamber which was present superior to the left of the enlarged LV. There was a well developed subaortic conus, which separated the aortic valve from the left A-V valve. VSD was subaortic and considerably large. The conal septum was not present.

In this case relation between the imaginary Sv-PA and Pv-AO lines were mildly spiral in counterclockwise fashion. The plane of the artificial conal septum was not concordant with the artificial sinus septal plane. They made approximately 60° on the left. An angulation of such degree could be adjustable by spirally formed artificial septum without inducing stenosis of the outflow tracts.

After enlargement of the VSD, the artificial septum was sutured the same way as in case 1. In this case a soft teflon patch (4 x 2 cm) had a long spade-like shape; The long leaf of the patch corresponding to the apical area, one of the short leaves to the sinus septum, and the other to the conal septum.

Intermitent coronary perfusion by releasing the aortic cross clamp was done to keep the heart in fibrillation state so that an appropriate patch size could be easily determined. The pulmonary band was not removed. After the procedure, EKG findings were the same as before surgery except increased QRS duration. Post-operative CHF was managed by anticongestive measures. Angiocardiograms performed 2 months after surgery demonstrated a moderate residual shunt. Pressures in the pulmonary and systemic ventricles were almost identical. It is now 9 months since the operation pt is doing reasonably well than before. Digitalis has been continued.

Case 3. A 10 year old female had signs and symtoms of large VSD. Ligation of a patent ductus anteriosus and a mild pulmonary artery banding had been performed previously.

Right heart catheterization disclosed left to right shunt at ventricular level with a ratio of 61%, and resistance ratio of 38%. The right ventricular systolic pressure was equal to that of the systemic.

Retrograde ventriculogram at what was thought to be left ventricle revealed simultaneous opacification of the aorta and hugely dilated pulmonary artery. Clinical diagnosis was VSD with a large left to right shunt and a mild pulmonary artery banding.

Cardiac Surgery was performed by Prof. Sakakibara. On inspection through the ventriculotomy, ventricular septum was virtually absent with normally interrelated great arteries (Type C I Solitus). The right and left A-V valves and their apparatus were well formed. The imaginary lines were parallel, the Sv-PA line being superior to the right of the Pv-Ao line. A straight artificial septum was laid between them.

Immediately after termination of the bypass, the systemic pressure was 120 mm Hg in systole, while that of the right ventricle 90 mm Hg. The post-operative EKG was completely the same as that before the surgery. The right heart catheterization at 3 weeks after the operation revealed a minor left to right shunt.

It is now 2 years after the surgery, the patient is doing quite well, and a recent chest x-ray revealed a normal cardiac size (CTR 53%).

DISCUSSION

There have been four reports dealing with this kind of intracardiac repair on single or common ventricle. The first report was by Sachs and associates (1968) in a 13 year old male with type C I Solitus and pulmonary stenosis. The second case was reported by Kawashima et al (1971) in a 8 year old female with type C I Solitus and pulmonary stenosis. The case 3 of this presentation has been published by Sakakibara et al (1972). To the authors' knowledge, these are the only successful cases in the literature.

Another case was reported by Horiuchi et al (1970) in a 6 year old girl with type A III solitus. She developed complete A-V block during the surgery and suddenly died while playing 4 months after the procedure.

Indications for the procedure: Relation between the two imaginary lines, morphologies of the A-V valves and their apparatus, size of the ventricular cavity, and associated anomalies of the outflow tracts and great arteries are the most important features in judging the indications.

Imaginary lines: The two imaginary lines (Sv-PA line and Pv-Ao line) should be parallel to construct a straight ventricular septum. Mild spiral formation less than 90° can be adjustable when placing the conal septum as in case 2. Such relation between the A-V valves and semilunar valves is present in i) A III solitus ii) A II Inversus iii) C III Solitus, iv) C II Inversus, v) A I Solitus vi) A iv Inversus, vii) C I Solitus and viii) C iv Inversus. Type A III solitus, ie single left ventricle with rudimentary outlet chamber, 1-transposition and situs solitus of viscera and atria is the most common type (40%), and especially those who had side by side great arteries as in case 1 are the most suitable candidates for this procedure.

A-V valve morphology: Only those who have well divided and well formed right and left A-V valves can be the candidates for this surgery. The papillary muscles of each A-V valves are preferably separated. Usually the atrial septum is well-formed in those cases who have well divided A-V valves.

A ventricular angiogram (preferably cineangio) is diagnostic in demonstrating well formed A-V valves by two negative jet during early diastole. Other valvular anomalies, such as stenosis, regurgitation should be evaluated.

Size of the ventricular cavity: From observations on these surgical cases and heart specimens with single or common ventricle, those who are more than five years of age appeared to have enough ventricular cavity size for the procedure. If the heart is small ventriculotomy becomes bigger in reverse proportion to the heart size, and there is a danger to induce stenosis of the newly made outflow tracts.

Removal of the supero-anterior rim of the bulboventricular foramen in type A single ventricle provides a wide outflow tract, and also may enlarge the newly made ventricle by incorporating the rudimentary outlet chamber. Many cases with left single ventricle have a sizable trabeculated sinus, which has been called "apical recess".

Associated anomalies: Less than moderate subaortic stenosis, which is usually due to obstructive bulboventricular foramen, can be amenable surgically, if it is not associated with critical hypoplasia or interruption of the aortic arch. As pulmonary valvular or subvalvular stenosis protects the pulmonary vascular bed from developing vascular obstructive disease, these are favorable unless it is not associated with severe hypoplasia of the pulmonary vascular tree.

If the patients have increased pulmonary blood flow a banding of the pulmonary artery should be performed during early infancy. This makes an intracardiac repair possible in later life as in case 2 and 3.

Surgical procedure: The most essential surgical aspects of this procedure are to avoid major conduction disturbances, equal partitioning of the uni-ventricular cavity without outflow tract stenosis, and adequate size of the artificial septum.

Conduction system in single or common ventricle has not been well-known. However, we can recognize where the atrioventricular node is, because in those cases with a normal atrial morphology, the atrioventricular node is always in the vestibulum of the coronary sinus. Another fact we know is that the major bundle branches are always directing toward the major papillary muscles. In single or common ventricle with well divided A-V valves, the coronary sinus is just above the posterior corner of the junction between the left and right A-V valves. Just below this posterior corner of the A-V valvular junction, so-called posterior median ridge is present in some cases as in case 1. It can be assumed that the His bundle may penetrate the fibrous tissue relating to this posterior junctional corner, and branches toward the major papillary muscles present. This should be demonstrated in serial sections of the specimens.

To avoid conduction injuries in the present cases, the artificial sinus septum was laid along the proximal portion of the medial leaflet of the right A-V valve until it passed the coronary sinus vestibulum, and then continued along the right side of the posterior wall far from the left papillary muscles. No critical conduction disturbances developed in the present series except mild increase of the QRS duration in case 1 and 2. This was probably due to injuries of the right side bundle branch by the sutures along the posterior free wall. As observed in case 3, it may be possible to avoid even this kind of minor injury, if interrupted sutures are employed. The posterior fixing line of the septum is far to the right in all present cases and completely dependent upon the position of the medial leaflet of the right A-V valve and also upon the relation between the left and right papillary muscles. In order to divide the univentricle into grossly equal proportions, the fixing line at the apical portion and the anterior free wall can be adjusted. In the post-operative angiocardiogram of the present series, the pulmonary venous ventricle is slightly larger than the systemic venous ventricle. Mild smallness of the systemic venous ventricle appears to be well tolerated.

Care should be paid not to induce outflow tract stenosis when inserting the canal septum. Enlargement of the VSD has been already mentioned.

If the created septum is excessive, it may induce inflow as well as outflow obstruction of the lower pressure ventricle. The size of the artificial septum should be just fit during the ventricular systole. A well calculated design of the artificial septum between the two imaginary line is necessary. In addition to this, in case 2, the heart was kept in fibrillation state by intermittent release of the aortic cross cramp during the septation along the apical and anterior free wall area, so that an appropriate patch size during systole could be assessed again. Until the organization of the artificial septum occurs, marked pressure difference between the newly made ventricles may be unfavorable. For this reason pulmonary artery banding of mild degree were not removed in case 2. It can be removed later. A soft Teflon patch is considered to be much more beneficial than a hard one, for this may not disturb ventricular contractility in any direction, and may easily adapt the appropriate shape after implantation.

Possibility: For those who have crossed imaginary lines, for example type C II solitus, A II solitus and so on, modified Rastelli's operation can be performed in selected cases. The sinus artificial septum is laid the same way as those with parallel imaginary lines, and the conal septal portion is so fixed as to connect both the great arteries with the newly made pulmonary venous ventricle. Then grafting between the systemic venous ventricle to the main pulmonary artery is performed utilizing a graft with a valve.

SUMMARY

A successful intracardiac repair with a straight artificial ventricular septum was presented on two patients with left single ventricle and 1-transposition of the great arteries, and on one patient with common or undivided ventricle.

The indications for this surgical procedure and future possibility on other types of single or common ventricle were discussed.