Complete atrioventricular septal defect: correction two patch technique

Komplet atriyoventriküler septal defekt: Düzeltme çift yama tekniği

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Background: Atrioventricular septal defects (AVSDs) include a variable spectrum of congenital malformations with different forms of clinical findings.

Methods: In this study, we report the surgical results for with this type of congenital cardiac malformation obtained in a single institution. Between November 1998 and January 2009, 58 patients with complete AVSD were operated on by the same team in the same department. In all patients, complete biventricular correction was performed. The mean bypass time (extracorporeal circulation; ECC) was 71.7 min. and the mean aortic clamping time was 50.1 min. The complete defects were corrected by the double patch technique and mitral cleft was closed in all except two patients.

Results: There was no intraoperative mortality, but hospital mortality was 6.9% (n=4), due to the pulmonary hypertension crises occurring in the first 15 postoperative days. During the follow-up four re-operations (6.9%) were performed: two for residual ventricular septal defects and two for mitral regurgitation. No mortality occurred during re-operations. In long-term follow-up mild and moderate -residual mitral regurgitation were detected in 26 and 3 patients, respectively.

Conclusion: Complete correction of AVSD can be carried out with acceptable results in a varied spectrum of anatomic forms and clinical severity despite the age of correction by using double patch technique.

Key words: Atrioventricular septal defects; mitral failure; pulmonary hypertension; surgical results.

Atrioventricular septal defects (AVSD) represent a full spectrum of lesions, characterized by the absence of the atrioventricular segment of the cardiac septum. As a consequence, the atrioventricular (AV) valves are malformed, mitral and tricuspid components are displaced at the same (Downward) level, the aortic root is not wedged, as is usual between the AV valves, but displaced forward, and the aortic valve profiles at a higher level. As a result, the left ventricular outflow tract is longer, narrower, and prone to obstruction - left ventricular outflow tract obstruction (LVOTO).

In complete forms there is a common AV ring and both valve components stay undivided. Anterior and posterior leaflets bridge the septum, and the anterior leaflet may adhere to the septal crest (type A of Rastelli classification) or float freely, straddling the septum, to
insert on right free wall (type B/C of Rastelli). The AV valve shows five leaflets, the mitral component being made of three leaflets - a small mural leaflet, an anterior and posterior leaflets, these being separated by a septal apposition area or cleft, an area that will normally leak, to produce mitral regurgitation. In a small percentage of cases the mitral component will show a single papillary muscle.

For complete defects, a large, nonrestrictive ventricular septal defects (VSD) and a primum atrial septal defect (ASD) are normally present, although the inlet area of the VSD usually presents numerous cordae, tethering valve tissue to the septal crest. Conduction tissue is abnormally placed in front of the coronary sinus, (sinus septum) and along the crest of the septum, until the level of the cleft, being more vulnerable to surgical injury.

Associated lesions are common: secundum ASD, persistent ductus arteriosus and left superior vena cava may all occur, but mitral regurgitation and dynamic pulmonary hypertension, mostly due to unrestricted left to right shunt are the rule. The association between AVSD and Down syndrome has long been known.

Complex forms of AVSD may indeed occur and include unbalanced ventricles, association with double outlet right ventricle and tetralogy of Fallot, but are be beyond the scope of this paper.

PATIENTS AND METHODS

Surgical indication and surgical objectives

The trend is for early primary correction, and most centers will presently recommend surgery between three and six months of age, the results being uniformly good. Banding of the pulmonary artery is exceptional and reserved for the unusual unbalanced forms or for severely unnourished babies. The objectives of surgery are:

- Complete separation of the circulations, by closing the VSD and ASDs
- Obtaining two AV valves with minimal or no regurgitation and no stenosis
- No heart block
- No subaortic stenosis
- Durable repair, without the need for late reoperations

These objectives may be achieved with different techniques - classical single patch technique, double patch technique and no VSD patch technique - all claiming equivalent results at present.

We will describe the technique we have been using for more than twenty-years now, with fairly reproducible results - the double patch technique.

Preoperative study

Patients are accepted on the basis of transthoracic 2 D-echocardiogram and the echo findings are confirmed by tranesosophageal echocardiography (TEE) on the operating table. Cardiac catheterization has became exceptional these days, and the minimal information we request from cardiologists is as follows:

- Confirmation of diagnosis - AV valves at same level and most AVSD features.
- Atrioventricular valve anatomy and function:
  - Common AV ring
  - Rastelli type (A/B-C)
  - Mitral valve regurgitation - amount and mechanism (cleft)
- Single papillary muscle?
- Ventricular septal defects size and morphology
- Atrial septal defect primum, secundum - size and morphology
- Balanced ventricles
- Exclude subaortic obstruction
- Exclude patent ductus and left superior vena cava

Only in late presenting cases, where pulmonary vascular disease is suspected, is a cardiac catheter requested.

Operation sequence

- Standard median sternotomy, partial resection of thymus and harvesting of a rectangle of pericardium (± 3x4 cm), to be pre-fixed in 0.6% glutaraldehyde for 10 minutes.
- Preparation for standard cardiopulmonary bypass, direct aorta and caval cannulation, bypass run at moderate hypothermia (30 °C) and at full flow. Aortic cross-clamping and cold blood cardioplegia, repeated at 20 minutes, plus topical cooling of the heart. Venting through the aortic root and modified ultrafiltration, for every case, after bypass termination.
- Patent ductus arteriosus (PDA) is ligated immediately at the beginning of the pump run, and the left superior cava cannulated directly, whenever present.
- The heart is approached by an oblique right atrial incision, prolonged anteriorly in front of the
inferior vena cava, for better exposure. Stay sutures are applied to the atrial wall, at 22/23 and 2/3 hours, evertting the AV valve ring. A sump sucker is introduced through the ASD secundum, to drain the left side.

- Anatomy is then inspected, confirming the echo findings; jet lesions, namely on the septal cleft, may suggest the regurgitation mechanism and a single papillary muscle must be excluded at this moment. The AV valve is than tested with saline, for competence and to find correction guidance points.

- If the defect is type A Rastelli, the centre of the valve over the septum is already defined for the anterior bridging component, and saline will only help to find the area where the posterior leaflet will make contact over the septal crest. If the defect is type C, the center of the valve, marking the septal origin of the cleft must be determined by trial and error. We always found that the most important defining suture is the one, first placed at the center of the future mitral valve (the extreme of the cleft, away from the septum).

To find that point we suggest:

- With a nerve hook, both the anterior and posterior leaflets on the left are sequentially explored, until the first primary mitral cordae is found. There, place the first suture (double armed nylon 5-0 or 6-0) to mark the center of the valve. The cleft is routinely closed by interrupted sutures, working from the center of the mitral component to the septal crest. This will make cleft closure simpler and mitral symmetry more easily achievable. Mitral valve diameter is now tested, with appropriate size Hegar dilators (Fig. 1).

- Whenever a single papillary muscle is found no cleft closure is done, as it would lead to mitral stenosis.

- Two stay sutures are now applied to the anterior and posterior bridging leaflets, on their right aspect, to expose the VSD. Ventricular septal defects patch is measured in length and high, leaving some 3 or 4 mm extra length posteriorly, and not making it too high or too short (risk for LVOTO). We normally use a patch of polytetrafluoroethylene (PTFE) material (0.1 mm) in a scooped rectangular shape. The patch is negotiated underneath the cordae, on the right aspect of the septum, some secondary cordae may have to be sacrificed. Suturing is continuous, using “CV 6 PTFE”, starting at the middle with a pledgeted suture, and running both ways to issue at the AV annulus; anteriorly where the atrial septum joins, and posteriorly, curving 3 to 4 mm to the right, to escape the conduction tissue area (Fig. 2).

- The ASD is now closed by using the autologous pericardial patch, that is trimmed at the table. The lower suture line is sandwiching the atrial

![Fig. 1. Marking sutures at the septal crest and the left atrioventricular valve component centre.](image1)

![Fig. 2. Measurements and tailoring the ventricular septal defect patch.](image2)
patch, the VSD patch and the valve tissue, and we tend to use interrupted “U” sutures (nylon 5-0 or 6-0), adapting for size. Once the valve level suture line is finished it is now time to finally check mitral valve competence with saline. Occasionally, additional central or commissural sutures may be needed to achieve competence (Fig 3).

- The atrial suture line is now brought, climbing the atrial wall, behind the coronary sinus (that we elect to leave on the left side, whenever there is no left cava), passing in front of the inferior cava, to meet the other suture arm, coming upwards from the left. We tend to close the secundum ASD and the primum defect with the same patch. The tricuspid valve is now tested with saline (Fig 4).
- The heart is de-aired and aortic clamp removed, the right atrium is closed routinely and weaning off bypass conducted as for any other case. Control TEE is repeated and pressures in the left atrium and pulmonary artery (PA) are measured for future reference. If PA pressure is found still high after filtration, and repair found to be adequate (no residual VSD, or mitral regurgitation, and no LVOTO), the patient will be considered as at risk for pulmonary hypertensive crisis in the Intensive Care Unit, and treated accordingly.
- Decannulation, heparin reversal and routine closure. No heart surface lines are left, only one atrial and one ventricle pacing wires.

RESULTS
Over the last nine years 58 babies with complete AVSD were consecutively corrected by one surgeon at our institution with the same team.

Results are presently excellent, with mortality rates below 7% and low reoperation needs, and survival time is long and free of any cardiac events. Surgical correction is being offered in early infancy, and with no added risk (Table 1).

DISCUSSION
Complete correction of complete AVSD can nowadays be achieved with low mortality and few complication rates. The trend is for very early correction, upon presentation.[1,2] The results have traditionally been dependent more on patient complexity than types of surgical correction used over the years. Complex forms, namely association with tetralogy of Fallot, double outlet right ventricle, unbalanced ventricular forms are associated to worse prognosis, and possibly severe mitral insufficiency. The classical association with Down syndrome seems to be more a protective element, than a risk factor and chromosomally normal children tend to have more dysplastic valves.[3]

As far as the surgical techniques are concerned, no differences were found between single patch, double patch,[4] or the newly introduced modified single patch technique.[5] However, closure of the
so called cleft (septal apposition area), whenever possible, seems now consensual and associated to less long time mitral regurgitation.\cite{6} The double patch technique is very reproducible for any type of lesions and for any age group\cite{1,7} and that is our experience, as well.

The use of the modified single patch repair, without the use of a VSD patch may not be applicable to large VSD components and might induce LVOTO for excessively lowering AV valve plane.

In conclusion, we present our results with the classical double patch technique that we have been using for decades, each surgeon should use the technique that he masters better, as all will give equivalent results. The cleft must be routinely closed, unless there is a single papillary muscle and repairs must be conducted at earlier stages before valve tissues get damaged by regurgitant jets, or pulmonary vascular diseases develop.

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