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Single-Stage Repair of Complete Atrioventricular Septal Defect

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Key-words: Total atrioventricular septal defect – AVSD – single-stage repair.

Schlüsselwörter: Kompletter atrioventrikulärer Septumdefekt – AVSD – einstufige Korrektur.

We report our experience with 31 consecutive children who underwent single-stage repair of complete atrioventricular septal defect between 1984 and December 1991. Ages ranged from 2 months to 2.5 years, mean 11 months. 18 were classified Rastelli type A, 13 type C. 22 patients had Down's syndrome, 12 were Rastelli type C. 1 patch was used in Rastelli type A cases and 2 patches in type C patients, without incision of the atrioventricular valve tissue. In all but 1 case the left superior and inferior valve leaflets were approximated. 5 patients died postoperatively resulting in an overall mortality of 16.1%. In all survivors, good clinical results and sinus rhythm were seen, although all show some degree of mitral incompetence.

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Einstufige Korrektur des kompletten atrioventrikulären Septumdefektes

Zusammenfassung: Wir berichten unsere Erfahrungen mit 31 Kindern, die im Alter von 2 Monaten bis 2,5 Jahren (im Mittel 11 Monate) zwischen 1984 und Dezember 1991 wegen eines kompletten atrioventrikulären Septumdefektes einer einstufigen Korrekturoperation unterzogen wurden. 18 Patienten wurden nach *Rastelli* als Typ A, 13 als Typ C klassifiziert. 22 Kinder hatten ein Down-Syndrom, davon waren 12 Rastelli Typ C. Beim Typ A wurde 1 Patch und beim Typ C 2 Patches ohne Inzision des atrioventrikulären Klappengewebes verwendet. Bis auf 1 Fall wurde bei allen Patienten das obere und untere linke Klappensegel adaptiert. Postoperativ starben 5 Kinder, entsprechend einer Gesamtmortalität von 16,1%. Alle Überlebenden sind im Sinusrhythmus und bieten gute klinische Ergebnisse, allerdings ist echokardiographisch bei allen eine Mitralklappeninsuffizienz nachweisbar.

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Introduction

Without surgical intervention, infants with complete atrioventricular septal defect (AVSD) have a poor prognosis: only about 35% survive the first year of life without operative treatment (2).

Historically, many surgical strategies have been used, in principle either as palliative or as primary corrective option. Single-stage complete repair will avoid immediate and long-term problems of palliative procedures as unpredictable functional results of pulmonary artery banding. Some other advantages include the avoidance of multiple surgical interventions with their cumulative morbidity and mortality, the elimination of the need for multiple and protracted hospitalizations, and the reduction of the psychological stresses imposed on the patient and family by the burden of a lingering cardiac lesion (1).

Since 1984 we therefore promote early repair without preceding palliation. The current study summarizes our surgical experience at the 2nd Surgical Department, University of Vienna, with a standardized technique for single-stage intracardiac repair of AVSD unaccompanied by other major cardiovascular malformations.

Patients and methods

From 1984 through December 1991 a total of 31 consecutive children have undergone single-stage repair of AVSD. Patients with other major malformations were excluded from this series.

Associated cardiovascular anomalies present in 14 children (45%) included patent ductus arteriosus (n = 8), atrial septal defect or patent foramen ovale (n = 5), mild infundibular pulmonary stenosis (n = 1), persisting left superior vena cava (n = 1), and coarctation of the aorta in 1 patient, treated 6 weeks before by subclavian-flap-technique and closure of the duct.

The age ranged from 2 months to 2.5 years (mean age 11.3 months) at the time of operation. The mean weight was 6.1 kg

(range 3.9 to 12 kg). During the period of 8 years in question, mean age and body weight at the time of operation decreased from 12.3 months and 6.3 kg during the first 4-years period to 10.3 months and 6.0 kg thereafter.

Classification of the defect was performed according to *Rastelli* (6): 18 defects (58%) corresponded with type A, 13 with type C. 22 patients (71%) showed Down's syndrome: 10 were of Rastelli type A, 12 of type C.

During the first years the diagnosis was verified by cardiac catheter. Since 1984, predominantly echocardiography was used, restricting catheterization for proving associated defects and for pressure measurements if considered necessary. In particular, ventricular function and incompetence of the mitral part of the atrioventricular valve were identified and delineated.

Progressive congestive heart failure with significant intracardiac shunting and pulmonary hypertension established the indication for a surgical procedure.

Operative technique

In most patients, the procedere was performed using cardiopulmonary bypass with moderate hypothermia to about 26°C. In 6 children deep hypothermia and circulatory arrest without cardioplegia was used after cardiopulmonary bypass-induced cooling to about 18 °C.

The surgical technique used was standardized: In Rastelli type A a single autologous pericardial patch is fixed with pledgeted interrupted sutures to the crest of the ventricular septum, thus even closing small defects at the base of the atrioventricular valve usually present. In the case of type C, the ventricular part of the defect is reconstructed with a semilunar Dacron patch which is sutured to the crest. Thereafter an autologous pericardial patch is connected to the Dacron patch with interrupted sutures incorporating the leaflets of the valve into the sutureline after having joined them together with a single suture close to the crest. In all but 1 case, the septal cleft was repaired with a few single sutures taking care not to create mitral valve obstruction. Finally, the atrial part of the defect is closed by the pericardial patch and continuous suture. In all patients this sutureline is placed such as to keep the orifice of the coronary sinus draining to the right atrium. This aim requires meticulous stitches to avoid surgically induced heart block. In patients undergoing operation in deep hypothermia, the period of cardiac arrest was between 41 and 66 min, mean 53 min. In all patients, aortic cross clamp time ranged from 45 to 87 min, mean 63 min. Postoperatively, all patients required mechanical ventilation and were treated with inotropic agents as required.

Since 2 years, transesophageal or epicardial echocardiography is used routinely to evaluate the functional result before leaving the operating theatre, as well as during the postoperative course.

Results

In this group of 31 consecutive patients undergoing single-stage repair of AVSD there were 4 deaths occurring during the perioperative in-hospital stay, one patient died early after discharge, thus adding up to a 30-day mortality of 16.1% (5/31). There was no late death.

One child with severe pulmonary hypertension (12.5 Wood units) died just 1 hour after the operation showing persisting right heart failure, another patient with pulmonary hypertension and chronic heart failure deteriorated progressively and died on postoperative day 3, one child finally deceased due to intractable severe mitral regurgitation. These 3 children were in normal sinus rhythm.

2 patients with sudden death, respectively sudden deterioration after a period of well-doing had a history of intraoperative total heart block turning into nodal rhythm or some atrioventricular dissociation alternating with sinus rhythm within a few days. One girl suffered sudden cardiac arrest minutes after having been transferred from the intensive care unit to the ward on postoperative day 4. The other child had already been discharged and was readmitted with signs of heart failure: after a few hours of clinical improvement he suddenly developed severe bradycardia, resuscitation failed in this case, too.

In the group of survivors, perioperative morbidity included mobile sternum (n = 1) necessitating reclosure of the chest, prolonged bleeding (n = 1) requiring reoperation the same day. Respiratory insufficiency, often combined with clinical signs of infection, was a common problem in the majority of patients, 4 developed prolonged pleural effusion.

In the follow-up reaching up to 8 years, all survivors have some degree of mitral insufficiency, though with wide range, minor residual leaks are known in 46% (12/26). Detail analysis of echocardiographic findings is a topic of a running study.

2 patients showed severe hemolytic anaemia, most probably caused by mitral insufficiency. At reoperation after 3 months, in 1 child both holes in the mitral leaflets and dehiscent sutures of the mitral cleft were found. Closure of the leaks and readaptation of the cleft eliminated hemolysis, a moderate regurgitation is still present. In the other child, most parts of the mitral leaflet were hypoplastic. This patient with additional appearance of ongoing sepsis is still ventilator dependent and in poor condition 2 weeks postoperatively.

After the operation had been postponed several times for various reasons, 1 child did not show any cardiac activity at the end of the procedure due to high potassium blood level caused by blood conserves prepared for an operation scheduled 3 weeks before. Fortunately, maximum output pacemaker stimulation was possible over the next 8 hours until normal potassium level was reached and sinus rhythm took over.

With exclusion of the patient with potassium-induced asystoly, 6 patients (20%, 6/30) had rhythm disorders intraoperatively (total heart block in 5, nodal rhythm in 1), requiring temporary pacing. 2 patients later on died, the remaining 4 have regained stable sinus rhythm.

Discussion

In this series, a standardized operative procedure has been choosen: for Rastelli type A a single-patch technique has been used, for type C the two-patch technique was the method of choice. In the latter case we thus avoid leaflet division, and it is our impression, that more accurate partitioning of the common atrioventricular valvar leaflet and easier restoring of valve function integrity is feasible. This experience corresponds with results reported by *Mavroudis* (4) and *Bove* (3) who were able to reduce mortality significantly using a two-patch-technique.

The operation is performed through a right transatrial approach providing excellent visualization and clear exposure of the common atrioventricular valve apparatus. This approach may even be improved by using deep hypothermia and cardiac arrest, but occasionally this will go hand in hand with some pressure due to restricted time available.

In our experience, good clinical results predominantly depend on successful restoring of left atrioventricular valve function, in addition ventricular and atrial septation without residual shunts will contribute. In this series we carried out the policy to repair the septal cleft with nonpledgeted sutures with taking care not to create mitral stenosis, but echocardiographically in most cases some degree of mitral regurgitation was present.

Even with an apparently technically satisfying operation major problems may occur: besides mitral valve incompetence

and significant residual shunting, hemodynamic embarrassment can be caused by preexisting pulmonary hypertension. Because of the possibility of development of pulmonary vascular obstructive disease, most patients will require surgical intervention during the first year of life (5) to avoid further complications.

In this small group of patients 2 patients died suddenly, respectively after some period of good postoperative recovery. Both had rhythm disorders, starting with complete heart block in theater, later changing into nodal rhythm or instable sinus rhythm alternating with atrioventricular dissociation. In both cases with otherwise unremarkable postoperative course we suggest that the rhythm disorder finally resulted in early death. As a consequence, decision for permanent pacemaker implantation should be handled more liberal.

The majority of patients were those with Down's syndrome. These children did not show an increased mortality, yet there was a tendency to a slightly longer period of ventilator dependency, mainly due to increased amount of secretion in the respiratory tract and more frequent signs of pulmonary infections. In general, patients with trisomy 21 usually had well-developed atrioventricular leaflets providing favourable conditions for the repair of the mitral valve, whereas some children without genetic defect presented with partially hypoplastic leaflets. According to the short time of follow up less than 8 years, no statement is possible concerning long-term results. So far, no further operation was required, especially no late mitral valve reconstruction or replacement or late pacemaker insertion. In conclusion, single-stage total correction in symptomatic children should be performed early in order to avoid pulmonary vascular complications. Comparably low risk supports the concept of primary repair. Above all, precise reconstruction of the mitral valve, avoidance of residual septal defects, and maintenance of sinus rhythm are essential for successful repair and good functional results.

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Postoperative Intensive Care in Infants and Children After Cardiac Surgery

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Key-words: Cardiac surgery – low cardiac output – intensive care – infants – clinical scoring systems.

Schlüsselwörter: Herzoperation – Herz-Kreislaufversagen – Intensivpflege – Kinder – klinische Scoresysteme.

Background: The postoperative course of infants and children after open heart surgery is often complicated by cardiopulmonary insufficiency or low cardiac output.

Methods: From January 1989 to April 1992 441 infants and children with congenital heart disease underwent cardiac surgery. 128 of these patients (29%) required prolonged or extensive intensive care because of cardiopulmonary insufficiency or low cardiac output. Aortic cross clamp and cardiopulmonary bypass times were measured in all patients. In the postoperative period duration of mechanical ventilation, duration of intensive care, special monitoring and therapeutic strategies and clinical scores were documented.

Results: The overall mortality rate was 9.9%, the mortality rate in patients with postoperative cardiopulmonary insufficiency or low cardiac output was 34%. The mortality rate increased significantly up to 73% when the cardiopulmonary bypass time exceeded 200 min. Mean duration of intensive care of survivors (S) and nonsurvivors (NS) was 10.3 ± 0.8 and 4.1 ± 1.2 days, respectively ($p < 0.01$), mean duration of mechanical ventilation was 7.1 ± 0.5 (S) and 4.1 ± 1.2 (NS) days, respectively ($p < 0.01$). NS had a significantly higher degree of physiologic derangement assessed by the Acute Physiologic Score for Children and needed more monitoring and therapeutic interventions assessed by the Therapeutic Intervention Scoring System than S.

Conclusion: Complex cardiac surgery, a cardiopulmonary bypass time over 200 min, high catecholamine infusion rates combined with a persisting low mean arterial pressure are associated with a high postoperative mortality rate in infants and children with congenital heart defects.

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Postoperative Intensivpflege bei Kindern mit angeborenen Herzfehlern

Zusammenfassung:

Grundlagen: Der postoperative Verlauf bei Kindern nach Operationen am offenen Herzen ist oft durch ein Herz-Kreislauf- bzw. Herz-Lungenversagen gekennzeichnet.

Methodik: Von Jänner 1989 bis April 1992 unterzogen sich 441 Kinder mit angeborenem Herzfehler einer Operation am offenen Herzen. Wegen eines postoperativen Herz-Lungenversagens benötigten 128 Kinder (29%) eine verlängerte bzw. besonders aufwendige Intensivpflege. Die Aortenabklemmzeit und die Herz-Lungenmaschinenzeit wurden bei allen Kindern angegeben. Postoperativ wurden die Beatmungsdauer, Dauer der Intensivpflege, spezielle Überwachungs- und Therapieformen und klinische Scores festgehalten.

Ergebnisse: Während die Gesamtmortalität 9,9% betrug, lag die Mortalität bei Kindern mit postoperativen Herz-Lungenversagen bei 34%. Bei langem Einsatz der Herz-Lungenmaschine (> 200 min) stieg die Mortalität bis auf 73% an. Die mittlere Dauer der postoperativen Intensivpflege betrug bei den Überlebenden 10,3 ± 0,8 Tage und bei den Nichtüberlebenden 4,1 ± 1,2 Tage ($p < 0,01$). Die mittlere postoperative Beatmung dauerte 7,1 ± 0,5 Tage bei den Überlebenden und 4,1 ± 1,2 Tage bei den Nichtüberlebenden ($p < 0,01$). Die Nichtüberlebenden benötigten einen deutlich größeren Therapieaufwand, gemessen am „Therapeutic Intervention Score", als die Überlebenden. Zusätzlich war der Schweregrad der Erkrankung, gemessen am „Akuten Physiologischen Score für

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