Irish Journal of Medical Science (1971 -) (2019) 188:475–479
https://doi.org/10.1007/s11845-018-1855-5

ORIGINAL ARTICLE

Long-term outcomes following partial atrioventricular septal defect (AVSD) repair in Ireland

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Received: 22 December 2017 / Accepted: 18 June 2018 / Published online: 25 June 2018

Abstract

Background We describe the long-term results of partial atrioventricular septal defect (AVSD) repair in a single centre encompassing a 22-year period. Described are rates of survival, reoperation and complications.

Methods We performed a retrospective review of 556 patients undergoing AVSD repair to identify the 51 patients who underwent partial AVSD repair in Our Lady's Children's Hospital, Crumlin, Ireland, between 1993 and 2015 with long-term follow-up where available.

Results A total of 29 (56.8%) of patients were male and mean age at operation was 3.32 years. Mean weight was 13.2 kg. Trisomy 21 was present in 29 (56.8%). Five patients (9.6%) had undergone prior surgery. Mean cardiopulmonary bypass time was 89 ± 36 min and mean aortic cross-clamp time was 57 ± 28 min. One patient underwent partial AVSD repair and concomitant tracheal resection and extracorporeal membrane oxygenation decannulation. One patient was managed with suture atrial septal defect (ASD) closure, the remainder with patch repair of ASD and mitral cleft closure. The length of hospital stay was 9 ± 5 days. Median follow-up was 6.06 years (IQR, 1.65–10.2 years). There were no early mortalities. One patient died 1 year following surgery (1.9%). One patient required reoperation at an interval of 2 years for severe mitral regurgitation (1.9%).

Conclusions Short- and long-term survival following partial AVSD repair in Ireland revealed excellent results compared with other published series. Reoperation incidence also compared excellently with other reports published in the literature.

Keywords Atrioventricular septal defect · Paediatric cardiac surgery · Repair

Introduction

Atrioventricular septal defects (AVSDs) encompass a spectrum of cardiac anomalies occurring from abnormal development of the endocardial cushions and resulting in a defect in the atrioventricular (AV) septum and AV valves [1]. The group encompasses 3% of all major congenital cardiac defects and lesions are classified into complete, transitional and partial [2]. A partial AVSD consists of a large ostium primum atrial septal defect (ASD) and a cleft between the left superior and inferior bridging leaflets of the atrioventricular valve. There are two distinct AV valve orifices corresponding to the mitral and tricuspid valves. Physiologically, this results in a shunt at the atrial level in addition to AV valve regurgitation. The degree of shunting is dependent on the ASD size and patients may remain asymptomatic for a number of years [1]. Treatment involves open via median sternotomy using cardiopulmonary bypass and the ideal timing of surgery is between 1 and 3 years of age [3].

There is a strong association between AVSD and trisomy 21 and approximately 50% of cases occur in patients with Down's syndrome [4, 5]. Whilst the incidence worldwide is approximately 1–2 per 1000 live births, Ireland has the highest rate of trisomy 21 in Europe at 1:546 live births [6]. As such, the number of patients undergoing AVSD repair is also increased relative to other European countries, making it a pathology frequently encountered by congenital surgeons in Ireland. The aim of this study was to perform a retrospective review of partial AVSD repair in Ireland over the past 20 years to delineate survival and re-intervention rate associated with the procedure.

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Methods

Ethical approval was sought and granted by the institutional ethics committee in Our Lady's Children's Hospital, Crumlin. A comprehensive retrospective review of patients undergoing partial AVSD repair was then carried out. All patients undergoing AVSD repair between 1993 and 2015 were identified from operative case logs. From a total of 556 patients with complete, transitional and partial AVSDs, we selected the 51 who underwent surgery for partial AVSD. No patient undergoing partial AVSD repair was excluded from this analysis. A comprehensive search of written patient charts, operation notes, echocardiographic studies, intensive care patient records, clinic notes and autopsy reports where applicable was carried out. Earlier records were handwritten whilst patients undergoing surgery more recently had digital operative notes, echocardiographic images and reports, intensive care notes and clinic letters. A database was compiled with the following datasets included for each patient: gender, date of surgery, date of birth, echocardiographic findings pre procedure, weight at operation, gestational term at birth (prematurity was defined as born before 37 weeks), presence of concomitant diagnoses, pre-operative status, prior surgery, surgeon, type of cardioplegia, cardiopulmonary and aortic cross-clamp times, minimum recorded temperature, operative findings and procedure, type of repair, patch materials, PDA ligation, post-operative rhythm, complications, need for reoperation, length of hospital stay, status at last follow-up and most recent echocardiographic findings. Regarding the extent of atrioventricular valve regurgitation, this was graded by a consultant cardiologist into none/trace, mild, moderate or severe based on subjective assessment of colour flow doppler appearance of the components of the MR jet from multiple views.

Data are presented as the mean ± standard deviation or frequencies as appropriate. Survival and freedom from reoperation were represented using Kaplan-Meier curves. Statistical analysis was performed using GraphPad Prism ver 7.0 for Mac OS X (GraphPad Prism Software Inc.).

Results

Pre-operative characteristics

A total of 556 patients underwent AVSD repair; of these, 51 patients underwent partial AVSD repair in Our Lady's Children's Hospital, Crumlin, between April 1993 and April 2015. Patient characteristics are summarised in Table 1. A total of 29 (56.8%) patients were male and 22 (43.1%) female. Mean age at operation was 3.32 years (range 0.12–14.5 years). Mean weight at operation was 13.2 kg (range 4.1–42.8 kg). A total of 42 (82.3%) were born between 37 and 40 weeks gestation, 5 (9.8%) were overdue by 2 weeks and 4 (7.8%) were premature. Of those patients born prematurely, the average gestational age was 32.5 weeks (range 29–36). Of these, one patient had a birthweight of 1.2 kg and had undergone a laparotomy and bowel resection prior to pAVSD repair. The same patient suffered a respiratory tract infection and developed rotavirus enteritis in the post-operative period. The same patient was well at his last follow-up 5 years post-operatively. Regarding syndromic abnormalities, 29 (56.8%) of patients had trisomy 21, one patient had Bardet-Biedel syndrome, cloaca and pulmonary hypoplasia and one patient had developmental delay and syndromic features with no specific diagnosis. Of the group, five (9.6%) had undergone prior surgery: two duodenal atresia repair, one morgagni hernia repair, one cholecystectomy, one gastrointestinal resection for necrotising enterocolitis (NEC) and one patient required extracorporeal membrane oxygenation (ECMO) prior to surgery due to concomitant tracheal ring stenosis.

Table 1 Pre-operative data for partial AVSD patients

Male	22 (43.1%)
Gestation	
Premature	9 (17.6%)
Term	37 (72.5%)
Overdue	5 (9.8%)
Weight at operation (mean ± SD)	13.2 ± 7.8 Kg
Age at operation (mean ± SD)	3.4 ± 3.5 years
Syndrome	
Trisomy 21	29 (56.8%)
Bardet-Biedel, cloaca, pulmonary hypoplasia	1 (1.9%)
Previous surgery	
Duodenal atresia repair	2 (3.9%)
Morgagni hernia repair	1 (1.9%)
Cholecystectomy	1 (1.9%)
GI resection for NEC	1 (1.9%)
Intubated	5 (9.8%)
ECMO	1(1.9%)
PDA	34 (66.6%)

SD, standard deviation; *Kg*, kilogram; *GI*, gastrointestinal; *NEC*, necrotising enterocolitis; *ECMO*, extracorporeal membrane oxygenation; *PDA*, patent ductus arteriosus

Intraoperative details

Five operating surgeons practiced in Crumlin during the study period in question with standardisation of operative practices within the institution. All procedures were performed via median sternotomy and with partial thymectomy. Continuous cardiopulmonary bypass was instituted via direct bicaval and aortic cannulation. If present, a patent ductus arteriosus (PDA) was ligated prior to application of the aortic cross-clamp (n = 34, 66.6%). Cold blood antegrade cardioplegia was administered to all patients at 20-min intervals in addition to topical

ice slush to ensure myocardial protection. Mean cardiopulmonary bypass time was 89 ± 36 min and mean aortic cross-clamp time was 57 ± 28 min. Minimum venous temperature was 30 ± 3.6 °C. As described above, one patient underwent concomitant tracheal resection and ECMO decannulation. One patient underwent partial AVSD repair and concomitant pectus excavatum correction using a pectus bar. Regarding partial AVSD technique, one patient was managed with suture closure of the ASD in addition to mitral cleft closure. The remainder underwent ASD patch repair with autologous pericardium ($n = 42$, 82.3%) or Dacron ($n = 5$, 9.8%). Three patients (5.8%) received pericardial patch closure of the primum ASD with a further suture closure of secundum ASD. All patients had suture closure of the mitral cleft and $n = 43$ (84%) patients were in normal sinus rhythm at the end of the case with no patient requiring a permanent pacemaker prior to discharge (Table 2).

Post-operative details

Average length of stay following surgery was 9 ± 5 days. No patient required a permanent pacemaker. Regarding complications, three patients developed post-operative chylothoraces requiring monogen and one patient developed a cerebrovascular accident as a result of embolic phenomenon intra-operatively (Table 3). Four patients developed lower respiratory tract infections requiring antibioics, four patients developed sternal wound infections requiring re-wiring or re-exploration under general anaesthesia and one patient was readmitted at an interval post surgery with a sternal wound infection requiring debridement and application of a vac dressing.

Follow-up and reoperation

Median follow-up was 6.06 years (IQR, 1.65–10.2 years). Two patients were lost to follow-up. There were no early mortalities (< 30 days). There has been one late mortality ($n = 1$, 1.9%) (Fig. 1). One patient whose post-operative recovery was complicated by respiratory tract infection and was discharged on day 12 post-operatively died of unknown causes a year following surgery. Regarding clinical and echocardiographic follow-up, for whom it was available, 11 (22.9%) had trivial or no MR, 28 (58.3%) had mild MR, four (8.3%) mild-moderate, four (8.3%) moderate and one (2%) patient severe (Table 3). This patient required a reoperation. Surgery was undertaken 2 years following the initial repair and encompassed mitral valve annuloplasty, and Alfieri stitch (Fig. 2).

Table 2 Operative details

Variable	*N* (%)
Operative findings	
Partial AVSD	44 (86%)
Partial AVSD double orifice LAVV	1 (1.9%)
Partial AVSD left SVC	3 (5.8%)
Partial AVSD, pectus excavatum	1 (1.9%)
Patrial AVSD, tracheal stenosis	1 (1.9%)
Partial AVSD, morgagni hernia	1 (1.9%)
Operative procedure	
ASD suture closure, mitral cleft closure	1 (1.9%)
Single patch repair, mitral cleft closure	44 (86%)
Single patch repair, mitral cleft closure, de vega annuloplasty	2 (3.9%)
Single patch repair, mitral cleft closure, pectus correction	1 (1.9%)
Single patch repair, mitral cleft closure, repair morgagni hernia	1 (1.9%)
Single patch repair, mitral cleft closure, tracheal resection, ECMO decannulation	1 (1.9%)
ASD patch type	
Pericardium	45 (90%)
Dacron	5 (10%)
PDA ligation	34 (66.6%)
Postop NSR	42 (82%)
Complete heart block	0 (0%)
Duration of CPB (min)	90 ± 36
X-clamp time (min)	57 ± 28

AVSD, atrioventricular septal defect: *SVC*, superior vena cava; *ASD*, atrial septal defect; *ECMO*, extracorporeal membrane oxygenation; *NSR*, normal sinus rhythm; *CPB*, cardiopulmonary bypass; *X-clamp*, cross-clamp

Table 3 Post-operative details

Variable	*N* (%)
Length of stay (days)	9 ± 5
Post-operative complications	
Chylothorax	3 (5.8)
CVA	1 (1.9)
LRTI	4 (7.8)
Sternal wound complications	4 (7.8)
Median follow-up (years)	6.06 (IQR 1.65–10.2)
Early mortality	0 (0%)
Late mortality	1 (1.9)
Mitral regurgitation at last follow-up	
Trivial	11 (22.9)
Mild	28 (58.3)
Mild-moderate	4 (8.3)
Moderate	4 (8.3)
Severe	1 (1.9)
Reoperation	1 (1.9)

Discussion

Since the first repair of an atrioventricular septal defect by Lillehei in 1955, there have been major advances in cardiopulmonary bypass, paediatric myocardial protection and surgical technique [7]. Those initial reports utilised cross circulation between patient and donor at the dawn of open-heart surgery and prior to the era of cardiopulmonary bypass [8]. Atrioventricular septal defects are frequently encountered in Ireland due in part to the high incidence of Down's syndrome in the country [6]. This study was undertaken to review the short- and long-term survival and reoperation rates following repair of partial AVSDs in a single institution.

No patient died within the first 30 days following surgery. This compares favorably to other series where early mortality rates range between 1.6 and 5% [9–12]. One of the largest series published by Najdawi et al. describes 334 consecutive patients with a 2% mortality rate at 30 days [10]. In a series of

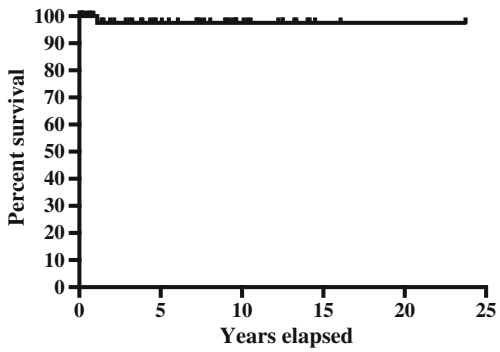

Fig. 1 Survival following partial AVSD repair in Our Lady's Children's Hospital, Crumlin, between 1993 and 2015

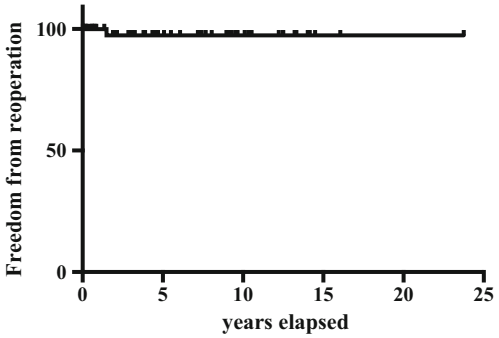

Fig. 2 Freedom from reoperation for mitral valve regurgitation following partial AVSD repair in Our Lady's Children's Hospital, Crumlin, between 1993 and 2015

133 patients described by Welke et al., mortality at 30 days was 5%; however, factors associated with mortality were not investigated by authors [11]. Burrato et al. report a 30-day mortality rate of 1.2% ($n = 3$) in their series of 249 patients [9]. There was one late mortality described in our series, which again compares favorably to previously published series [10, 11].

Regarding the incidence of complications, no patient in our series required a permanent pacemaker; this compares favorably to previous publications where the pacemaker rate cited is typically around 3% [9, 10]. Chylothorax following cardiac surgery occurs in 1.5–7.6% of all paediatric cardiac surgery cases and is associated with significantly longer duration of mechanical ventilation, hospital stay and in-hospital mortality [13]. This is unlikely to have impacted upon early mortality in our series, as this was zero. However, our rate of chylothorax was at the upper end of the spectrum for paediatric cardiac surgery patients at 5.9%, and this is possibly due to the high proportion of trisomy 21 patients in our cohort, which is known to be an independent risk factor for its development [14]. All three patients in our series developing chylothorax did have trisomy 21. The incidence of cerebrovascular accident (CVA) associated with partial AVSD repair in our centre during this period was 1.9%. In a large study undertaken to identify incidence of CVA amongst children with congenital heart disease undergoing cardiac surgery, the frequency of vaso-occlusive stroke was found to be 5.4 cases per 1000 children [15]. However, the case in question was particularly complex and was the first reported case of a patient requiring ECMO for acute respiratory distress resultant from tracheal stenosis for whom a slide tracheoplasty was successfully undertaken in conjunction with repair of the partial AVSD [16]. Furthermore, the patient made a full recovery from any neurological deficit experienced.

The reoperation rate described in our cohort was 1.9% and described one case requiring mitral valve annuloplasty and Alfieri stitch to achieve mitral competence 2 years after the initial repair. Again, this compares favorably to other series, which quote an 11–16% reoperation rate [9, 10].

Conclusion

Short- and long-term survival following partial AVSD repair in Ireland revealed excellent results compared with other published series. Reoperation incidence also compared excellently with other reports published in the literature.

Compliance with ethical standards

Ethical approval was sought and granted by the institutional ethics committee in Our Lady's Children's Hospital, Crumlin.

Conflict of interest The authors declare that they have no conflicts of interest.

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