

Improvement after Surgical Closure of Secundum Atrial Septal Defects in Adults

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Abstract

Background: Atrial septal defect (ASD) is the most common congenital heart disease (CHD) in adults after bicuspid aortic valve. Although the defect is often asymptomatic until adulthood, undetected ASDs could lead to potential irreversible complications like arrhythmias, stroke, pulmonary hypertension and its sequel.

The purpose of our study was to determine the value of surgical closure of atrial septal defects in adults and the effect of age on the prognosis of these patients.

Methods and patients: Retrospective study was carried out on 489 patients with age 18 to 65 years old who underwent surgical repair of isolated Secundum type ASD. There were 318 women and 171 men (mean age was 25.21 ± 10.106 years). The patients were divided into two groups, group I: from 18-40 years old. Group II: 40-65 years old. Preoperative, operative, and postoperative data were reviewed. Statistical analysis was performed.

Results: A statistically significant improvement of NYHA Functional class, tricuspid valve regurgitation and pulmonary artery pressure in most of the patients after operation. No residual intra-cardiac shunt was identified on echocardiographic follow up. Postoperative morbidity and mortality were higher in the older patients with high pulmonary pressure.

Conclusion: Surgery for ASD in the adult age is a safe, beneficial, and a low-risk option that modifies the patient's natural history by improving their clinical status. The operation must be performed without major delay, better to be in established GUCH centers by well trained staff. Despite the use of sildenafil is still need more research. It seems to have good result post-operative.

Keywords: ASD; Adult; Closure; Pulmonary artery pressure

Abbreviations: ASD: Atrial Septal Defect; CHD: Congenital Heart Disease; GUCH: Grown-up Congenital Heart Disease; TTE: TRANS-Thoracic Echocardiography; TEE: Trans-Esophageal Echocardiography; NYHA: New York Heart Association; VSD: Ventricular Septal Defect; BMI: Body Mass Index; AF: Atrial Fibrillation; PAP: Pulmonary Artery Pressure; SAID: Non-Steroidal Anti Inflammatory Drugs; ARDS: Adult Respiratory Distress Syndrome; SVT: Supraventricular Tachycardia

Introduction

Patients with congenital heart diseases, especially those with low mortality in childhood, are more frequently reaching adult age. Isolated Secundum type atrial septal defect (ASD) is the most common form of congenital heart disease presented in adulthood after bicuspid aortic valve and mitral valve prolapse [1,2]. Pediatric cardiac surgery is an important landmark in the history of patients with CHD, since, before its development less than 20% of these children could survive to adult age [3]. Now we know that the majority of deaths occur at this

adult age is greater than that of children with CHD [4,5]. These (GUCH) patients need special attention in an adequate setting in order to manage their problems and subsequent events intrinsic to the heart defect and also regarding age related co-morbidities [6].

There are several reasons, why patients may present with uncorrected lesions: late diagnosis is always a possibility, particularly in the case of ASD and aortic coarctation, as it is often clinically silent. Also other cases knew the lesion, but still reluctant or worried about the surgery, or those who need re-operations after successful repair because of residual defects or complications [7]. The progression of this congenital defect to congestive cardiac failure follows the onset of pulmonary hypertension, arrhythmias, respiratory infections, and other cardiovascular disease. Hence, the defect is usually discovered when a patient presents with dyspnea or palpitations, or occasionally on routine medical examination [8,9].

The main indication for ASD closure is the presence of a significant shunt as evidenced by right heart volume overload with or without symptoms (exercise intolerance, fatigue, dyspnea, heart failure, paradoxical emboli, and arrhythmias) [10,11]. Surgical repair of ostium Secundum or sinus venosus ASD performed early in childhood

without significant residual lesions could be considered effective surgery [12]. The closure of an ASD in an adult, even when there is a significant left to right short circuit, may be less than satisfactory, and there is still doubt if these patients will benefit from surgical closure of atrial septal defects [13-15].

The purpose of our study was to determine the value of surgical closure of atrial septal defects in adults and the prognosis of these patients in relation to age. Also the impact of sildenafil use in pulmonary hypertensive cases.

Methods and Patients

Retrospective study was carried out on 489 patients with age 18 to 65 years old who had undergone surgical repair of isolated secundum type ASD. The study was in 3 tertiary centers in developing countries, between 2010 to 2014 and was approved by ethical committee of the tertiary centers. All clinical data and investigations concerned for them were collected and analyzed pre and post-operative (clinical, demographic, radiographic, electrocardiographic, echocardiographic (TTE) and (TEE) if done. All data were collected from hospital files after the consent was taken from the patients and near relatives. Inclusion criteria: All cases \geq 18 years (adult congenital heart disease), with isolated secundum ASD type. Patients with normal PAP and those with reversible PAP by its criteria. Also, cases with sever PAP who received Sildenafil preoperatively.

Exclusion criteria: patients below 18 years old, those with other associated congenital heart diseases or acquired heart diseases. Cases with other types of ASD e.g. sinus venosus defect, ostium primum defect or unroofed coronary sinus. Patients with irreversible pulmonary hypertension confirmed by right heart catheterization.

There were 318 women (65%) and 171 men (35%) (Mean age: 25.21 \pm 10.106 years). The patients were divided into two groups, group I: from 18-40 years old and Group II: from 40-65 years old. The diagnosis was confirmed by trans-thoracic echocardiography (TTE). The use of trans-esophageal echocardiography (TEE) was established in poorly visualized cases with routine TTE. Pre-operative and post-operative follow up was established with consultant cardiologists. Coronary angiography was performed for all patients over 40 years of age at presentation. Right heart catheterization was performed in all cases with sever pulmonary hypertension (SPAP above 50 mmHg). Post-operative echocardiography was done for all cases before discharge and 12 months post-surgery.

Surgical procedure

All operations were performed on pump using heart lung machine with moderate hypothermia around 32°C. The chest was opened through a median sternotomy or right thoracotomy. After total cardiopulmonary bypass had been instituted, cold crystalloid cardioplegic solution (St Thomas' Hospital formula) was given antegrade through the aortic root in 290 cases and Fibrillator was used in 199.

In our study the method used to close the defect depended on its size. 154 patients with an isolated ASD underwent direct suture closure. The remainder required patch closure (with autologous pericardium or bovine pericardial patch) with either single or double layers of continuous sutures using Prolene 5-0. After weaning off bypass they were shifted to post-operative care unit for follow up then to regular ward till discharge home. Clinical data as New York Heart

Association (NYHA) functional class and echocardiographic follow up evaluations was performed for all patients at 12 months intervals, or as soon as either new symptoms appeared or the previous clinical state deteriorated. All patients were followed up for 1 year. We compared clinical data, functional capacity, rhythm status, and echocardiographic parameters of all patients before and after the operation in both groups.

Statistical analysis

All data were collected, organized and statistically analyzed using SPSS software statistical computer package version 13 (SPSS, Chicago, Illinois, USA). For quantitative data, the range, mean, and standard deviation were calculated. Correlation between variables was evaluated. Significance was adopted at $P < 0.05$ for interpretation of results of tests of significance.

Results

There were 489 patients, 318 women (65%) and 171 men (35%), who met the inclusion criteria made up the population for this study. Mean age was 25.21 \pm 10.106 years, with a range of 18 to 65 years of age. At the time of clinical evaluation, all patients were non cyanotic at rest and during exercise. 53 patients did not have significant symptoms, they were accidentally discovered. Symptoms as exercise intolerance, dyspnea or easy fatigability were the most significant finding in our patients. (Table 1)

Manifestations	Group I (292 cases)		Group II (197 cases)		p-value
	Number	%	Number	%	
Asymptomatic	18	6.2	35	17.8	Ns
Dysnea	82	28.08	97	49.2	<0.05*
Palpitation	125	42.8	105	53.3	<0.05*
Congestive symptoms	44	15.1	42	21.3	Ns
Stroke	12	4.2	6	3.1	Ns
Liver dysfunction and coagulopathy	13	4.4	9	4.5	Ns

Table 1: Presentations of cases in both groups. *P value<0.05 significant, Ns = Non-Significant.

Pre-operative clinical status and presentations were variable within each group of cases, but showed no significant difference between both groups (Table 1).

Palpitation was found in 125 cases (42.8%) in group I and 105 cases (53.3%) in group II. normal cardiac rhythm was found in 259 patients (52.9%). Tachyarrhythmia causing palpitation was found in 230 patients (47.1%) in both groups. They were divided by ECG into 55 cases with supraventricular tachycardia and 175 cases with atrial fibrillation. In the post-operative follow up, there was significant reduction of cases to 73 cases 14.9% with remained AF in a rate ranging between 75-110 beat/min under control using B blocker. Syncope was happened in 21 cases, secondary to rapid atrial fibrillation and supraventricular tachycardia.

Regarding systemic congestive symptoms, there was no significant difference between both groups. They were found in 44 cases in group I

and 42 cases in group II .18 patients were presented after paradoxical embolization causing stroke with 13 cases in group I and 9 cases in group II (Table 1).

In this study NYHA class II was 83 cases (28.4%) in group I and was 39 cases (19.8%) in group II. In group I NYHA class IV was 25 cases (8.6%) and in group II NYHA class IV was 42 cases (21.4 %). We noticed that, NYHA class III and IV were 27.9% and 21.4% respectively in group II of patient which were higher than the group I. There was significant correlation between NYHA class preoperatively and age of the patients as NYHA class preoperatively more higher in group II than group I (Table 2).

NYH A	Group I (18-40 years) 292 cases		Group II (40-65 years) 197 cases	
	Pre-operative	Post-operative	Preoperative	Post-operative
	N (%)	N (%)	N (%)	N (%)
I	127 (43.5%)	215 (73.6%)*	61 (30.9%)	113 (57.4%)*
II	83 (28.4%)	68 (23.3%)*	39 (19.8%)	74 (37.5%)*
III	57 (19.5%)	9 (3.1%)	55 (27.9%)	8 (4.1%)
IV	25 (8.6%)	0 (00%)	42 (21.4%)	0 (00%)
Total	292 (100%)	291(99.7%)	197 (100%)	195 (98.9%)

Table 2: Functional status (NYHA class) of patients pre-operatively and post operatively for both groups. *Significant (p<0.05).

NYHA class II, III and IV were significant finding in 122, 112 and 68 patients respectively preoperative in all cases (Table 3).

NYHA	Pre operative		Post operative	
	N	%	N	%
I	188	38.5	327	67.3*
II	122	24.9	142	29.2
III	112	22.9	17	3.5
IV	68	13.7	0	00.0
Total	489	100	486	100

Table 3: Improvement in NYHA class post-operatively as a whole. *Significant (P<0.05).

Regarding the surgical incision used, 228 cases underwent median sternotomy in group I and 154 cases in group II. On the other hand 64 cases underwent thoracotomy in group I and 43 cases in group II (Table 4).

	Group I (292 cases)		Group II (197 cases)	
	Number	%	Number	%
Sternotomy	228	78.1	154	78.2
Thoracotomy	64	21.9	43	21.2
Cardioplegia	185	63.4	105	53.3

Fibrillator	107	36.4	92	46.7
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Table 4: The type of incision and use of cardioplegia.

Degree of regurge	1 st group (18-40 years) 292 cases		2 nd group (40-65 years) 197 cases	
	Pre-operative	Post-operative	pre op.	Post-operative
	N (%)	N (%)	N (%)	N (%)
No regurge	11 (3.8%)	118 (40.4%) *	10 (5.1%)	80 (40.6%) *
Mild	113 (38.8%)	173 (59.2%) *	61 (30.9%)	115 (58.4%) *
Moderate	108 (36.9%)	0 (00%)	117 (59.4%)	0 (00%)
Sever	60 (20.5%)	0 (00%)	9 (4%)	0 (00%)
Total	292 (100%)	291 (99.7%)	197 (100%)	195 (98.9%)

Table 5: Tricuspid valve regurgitation preoperative and postoperative. *Significant (p<0.05) improvement in tricuspid regurge.

In this study cardioplegia was given to 185 cases in group I and to 105 cases in group II. Fibrillator was used to fibrillate the heart in 107 cases in group I and in 92 cases in group II. No significant surgical technical difference for those cases either sternotomy or thoracotomy or according to the method used to arrest the heart (Table 4). No residual intra-cardiac shunt was identified on echocardiographic follow up.

Overall patients were followed up for 1 year. There were significant improvement of NYHA functional class, tricuspid valve regurgitation and pulmonary artery pressure in most of the patients after operation with more improvement in group I than group II (Tables 2 and 3). Cases in our study with pre-operative tricuspid regurgitation showed significant improvement post-operatively in group I more than in group II (Figure 1). There was no tricuspid regurgitation in 118 cases (40.4%) post-operatively in group I and in 80 cases (40.6%) in group II (Table 5).

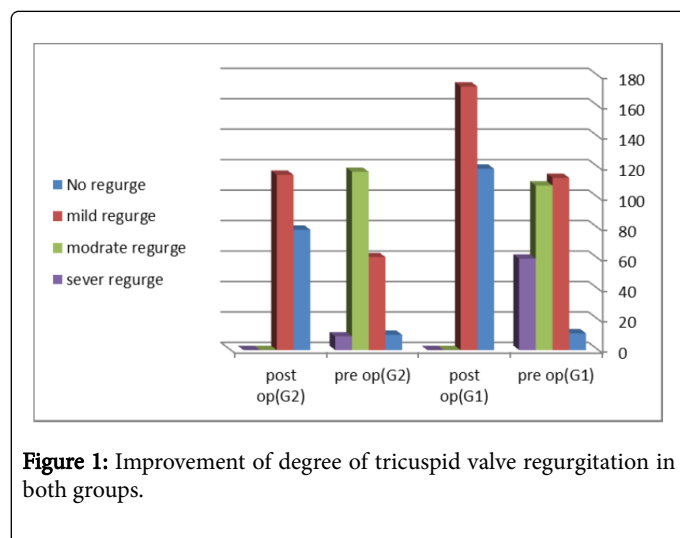


Figure 1: Improvement of degree of tricuspid valve regurgitation in both groups.

On measuring PAP post-operatively, There was significant reduction of it to the mean 14.9 ± 14 mm Hg, (minimum pressure was 5 mmHg and maximum was 30 mmHg), from pre-operative mean PAP 55 ± 13 mmHg in group I. Also in the group II, Mean PAP pre-operatively was 43.23 mmHg (minimum pressure of 26 mmHg and the maximum pressure was 99 mmHg) and significantly declined to be post-operatively mean PAP was 23.9 ± 15 mmHg, (minimum pressure was 10 mmHg and the maximum pressure was 45 mmHg). Over all pulmonary artery pressure showed significant reduction in our cases post-operatively with group I more than in group II (Table 6).

Pulmonary pressure	1 st group (18-40 years) 292 cases				2 nd group (40-65 years) 197 cases			
	Pre-operative		Post-operative		Pre-operative		Post-operative	
	N	%	N	%	N	%	N	%
<40	114	39	250	85.6*	62	31.5	160	81.2*
≥ 40-50	125	42.8	42	14.4	84	42.6	31	15.7
≥ 50	53	18.2	0	0	51	25.9	4	2.03
Total	292	100	291	99.7	197	100	195	98.9

Table 6: Pulmonary artery pressure by echocardiography. *Significant ($p < 0.05$).

There was significant correlation between age of patients and hospital stay, as it was shorter in group I than the group II. In group II some complications occurred and these complications were the leading causes to prolong their hospital stay (Table 7).

Variables	Group I (18-40 years) 292 cases	Group II (40-65 years) 197 cases	p value
Hospital stay (days)	10.69 ± 0.5	15 ± 1.5	$p < 0.05^*$
ICU stay (days)	2.63 ± 1	3.04 ± 0.5	Ns
Ventilation hours	13.75 ± 2	15.27 ± 3	Ns
Postoperative bleeding	7 cases (2.4%)	31 cases (15.7%)	$p < 0.05^*$
Pericardial effusion	19 cases (6.5%)	60 cases (30.4%)	$p < 0.05^*$
Wound infection	13 cases (4.5%)	28 cases (14.2%)	$p < 0.05^*$
Mortality	1 (0.3%)	2 cases (1.01%)	Ns

Table 7: Post-operative data in the two groups of patients including mortality. *Significant ($p < 0.05$).

Regarding the post-operative morbidities, there was significant correlation between age of patients and post-operative morbidities as wound infection, pericardial effusion and bleeding secondary to coagulopathy with higher occurrence of these complications in the group II of patients. The postoperative medical bleeding occurred in 7 cases (2.4%) in group I and in 31 cases (15.7%) in group II. The postoperative pericardial effusion occurred in 19 cases (6.5 %) in group I and 60 cases (30.4%) in group II the majority managed with

diuretics, NSAID and follow up. The postoperative wound infection found in 13 cases (4.5%) in group I and 28 cases (14.2%) in group II. There was no significant difference between both groups regarding ICU stay and ventilation hours (Table 7).

Our study showed that, the mortality was documented in group I and II. There were 3 deaths in the series. First case was a 49 years old from group II female died intra-operatively most probably due to irreversible protamine hypersensitivity, the second case was a 33 years old male died at the 4th post-operative day most probably due to ARDS, and the third post-operative death was a 65 years old male patient who died in the second post-operative day due to massive cerebrovascular accident (Table 7).

In our study, there was significant improvement in the quality of life as improvement of symptoms as dyspnea, palpitation physical activity and congestive manifestations (Figure 2). Also we can notice significant improvement post operatively in group I and II, but more in group I (Figure 3).

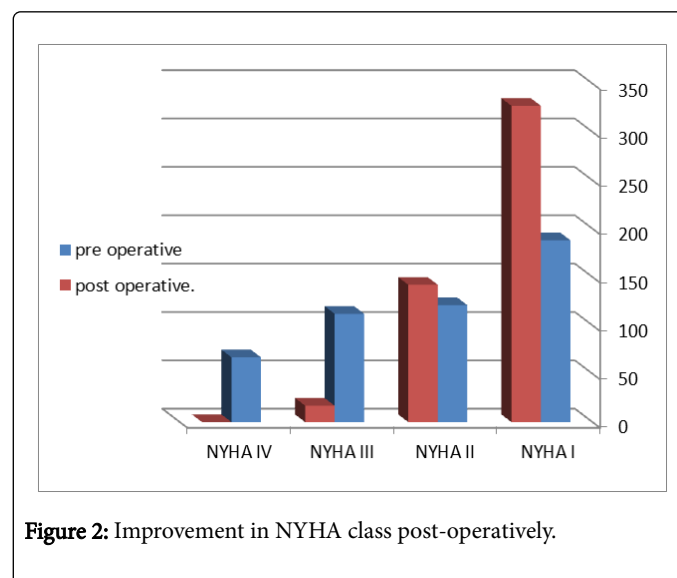


Figure 2: Improvement in NYHA class post-operatively.

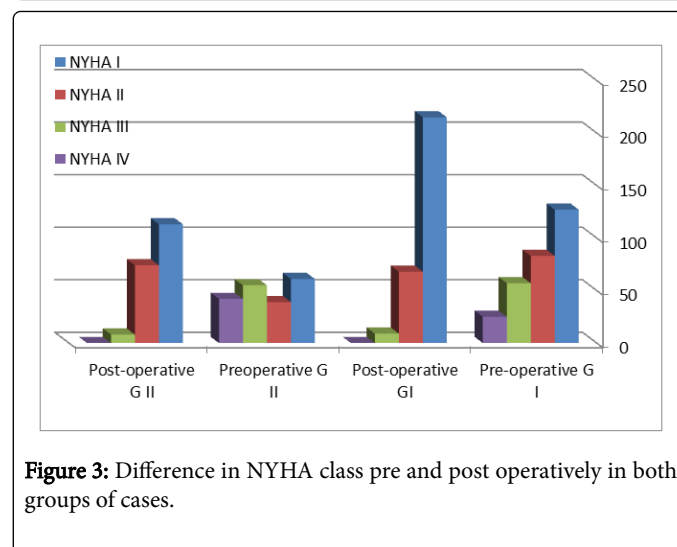


Figure 3: Difference in NYHA class pre and post operatively in both groups of cases.

In this study, sildenafil therapy showed significant improvement of PAP postoperatively. Cases with significant pulmonary hypertension (SPAP above 50 mmHg) were 104 cases; they received sildenafil for 6

months preoperative. In group I about 53 cases had severe pulmonary hypertension and 51 cases in group II preoperatively. In the postoperative follow up there was high significant improvement in PAP in patients who received sildenafil in both groups, as only 4 cases in group II still had high PAP (Table 8).

PAP	1 st group (18-40 years) 53 cases				2 nd group (40-65 years) 51 cases			
	Pre-operative		Post-operative		Pre-operative		Post-operative	
	N	%	N	%	N	%	N	%
≥ 50 mmHg	53	100	0	0	51	100	4	

Table 8: Post-operative PAP after the use of Sildenafil in 104 pulmonary hypertensive cases pre-operatively.

Discussion

There is no precise data about the group of grown-up congenital heart disease patients (GUCH) in the Middle East. This needs multicentre collaboration. In Middle East countries, these cases diagnosed late either due to lack of facilities or accidentally discovered recently with routine investigations. Also, certain cultures, still worry to do open heart surgeries in young age for their children. Our study doesn't provide definitive data about the prevalence of congenital heart disease (CHD) in adults; it gives an indirect picture of CHD in adult patients and its correlation with age.

In one of studies which were carried out on 2004 concerned in the follow up of cases, they found the predominated GUCH cases were VSD 31% and ASD 29%. Pulmonary and aortic stenosis, aortic coarctation, and complex congenital heart disease were less frequent. 75% of these cases presented with small lesions, which permitted the normal development of the patient and did not require specific therapy, 17% patients presented lesions of considerable impact and were waiting for cardiovascular intervention at the time of the study [12].

Because of that, we were concerned with the ASD cases as one of the predominant congenital cardiac lesion reaching adulthood. In our study some of the patients were asymptomatic and can survive easily to adulthood, unless, there is an associated lesion, or the balance between pulmonary and systemic circulation start to disturb. This was supported by many literature reviews, as ASD is the commonest congenital heart anomaly that can be seen in the adult age and secundum type is the most presented type of ASD. A physical examination of a patient with ASD usually reveals subtle findings, and hence the diagnosis may be missed [16].

The diagnosis was established by TTE in the majority of cases and only TEE was requested to confirm the diagnosis as they were of high BMI more than 35. Other studies confirmed the use of echocardiography for diagnosis as standard technique. Cardiac catheterization is wasn't necessary to complete the diagnosis but remains a critical technique for evaluation of irreversible pulmonary hypertension or for detecting underlying coronary artery atherosclerosis and for planning interventions, this is supported by other studies for adult CHD [17,18].

The use of coronary angiography was performed on all patients over 40 years of age and high risk cases for ischemic heart disease. Right

heart catheterization were done for cases with severe pulmonary hypertension more than 50 mmHg to exclude cases with irreversible pulmonary vascular resistant. Other investigators found 4% of cases had Eisenmenger syndrome secondary to the congenital heart defect, which rendered surgery impossible, whereas 4% had a significant lesion and rejected the surgery [12].

In this study there were variable presentations at the time of examination pre-operative, the most significant manifestations were palpitation and dyspnea and their impact on exercise intolerance and easy fatigability. The occurrence of stroke secondary to paradoxical emboli was documented in our cases encouraged surgery to avoid repeated cerebrovascular accidents. The findings were similar to other studies concerned in these symptoms and concluded that, the indications for closure in the pediatric and the adult population is essentially the same. In pediatric patients, however, primary attention is directed to symptomatology of recurrent respiratory tract infections and failure to thrive. In adults, respiratory symptoms such as shortness of breath tend to occur [19,20].

Classic sternotomy still the traditional approach to the heart, it was used in the majority of cases in both groups of cases, as most of the surgeons are not familiar with minimal invasive approaches either due to insufficient training or lack of facilities. Thoracotomy was used to less extent in cases of both groups in this study. Thoracotomy cases showed better recovery, more cosmetic satisfaction and decreasing the risk of sternal wound infection and sternotomy pain. The use of Fibrillator was used in a good number of cases in both groups comparable to the use of routine cardioplegic solution. In other study, all patients either post sternotomy or thoracotomy recovered rapidly from the surgery. Follow-up was complete in all patients, with no late complications and no residual shunt post ASD repair [21].

Regarding palpitation secondary to AF or SVT, was sharply decline post-surgical repair, confirmed with ECG. This coincides with the results of some authors who mentioned that surgical repair of ASD is a useful treatment in adult patients, because it improves hemodynamic status and normalize PAP, independent of age at the time of repair [22].

Pulmonary arterial hypertension, defined as a mean pulmonary arterial pressure greater than 25 mmHg at rest or greater than 30 mmHg during exercise [23]. According to this definition, most of our patients have pulmonary hypertension at presentation, Pulmonary artery pressure showed significant reduction post-operatively in both groups of cases with valuable reduction in group I, denoting correlation between age and PAP improvement post ASD closure. Some studies demonstrated in their work, 10% to 41.8% of patients developed pulmonary hypertension in varying degrees in adults with congenital heart diseases [8,23,24]. Also, an interesting reference in the work of some authors, who supported that in a number of adults even with severe hypertension pre-operatively, had a good post-operative course [25].

The principal consequence of ASD is a volumetric overload of the small annuli that causes progressive dilatation of the right ventricle and increases pulmonary pressure. These hemodynamic effects may be well tolerated for decades, but eventually cause right ventricular dysfunction and cardiac insufficiency. Tricuspid valve insufficiency secondary to annular dilatation is produced, that aggravate the sequences of ASD in adults [26]. Based on that Echocardiography was beneficial in follow up of cases pre and post-operative and confirmed the significant regression in the degree of tricuspid valve regurgitation.

When the ASD is closed before the age of 18 years, the majority of these changes normalize [12,27]. This coincides with our results that encourage surgery without delay.

In our study the morbidities as bleeding, pericardial effusion and wound infection were documented in both groups and their percentage were more in group II than group I. This correlations between the morbidities and age of patients were documented in other literatures concerned in post-operative complications [28].

Mortality was low among our cases as it was 0.61%, deaths occurred in 2 cases in group II and one case in the group I. Some authors estimated the mortality in their study, it was 3.3% all of them were in the older age group as well [14]. The mortality was much less with other research studies in young patients [29,30].

Throughout the 40 years over which surgical correction has been carried out, peri-operative mortality has decreased from 12.5% in the late 1950 s to 6% in the 1960 s, and to below 0.5% today [30]. This major improvement in survival appears to be a reflection of both better operative technique and better post-operative care, so surgical closure of this relatively common anomaly became widely recommended even for older patients.

Surgeries were done in these adult cases once diagnosed and according to pre-operative evaluation. This is confirmed with others who suggested that, surgery should be performed in the younger patient and probably before structural changes in the myocardium or pulmonary vasculature may start [31]. Surgery performed in our cases in both groups of cases after exclusion of irreversible pulmonary hypertension and this is supported by others who found that, age was a significant and independent predictor of surgical mortality and morbidity at late follow up [13].

Regarding the use of sildenafil, it showed significant improvement in PAP post-operative in both groups who received the medication. Literatures concerned about sildenafil found that, mid-term oral sildenafil therapy led to significant improvement in pulmonary hemodynamics, functional class and/or exercise tolerance [32,33]. Its use still need work up to prove its efficacy in pulmonary hypertension secondary to congenital heart defects.

Conclusion

Surgery for ASD in the adult age is a safe, beneficial, and a low-risk option that modifies the patient's natural history by improving their clinical status. The operation must be performed without major delay, better to be in established GUCH centers by well trained staff. Despite the use of sildenafil is still need more research. It seems to have good result post-operative.

Declarations

Consent for publication

Written informed consent was obtained from the patients for publication of this study. The consents are available for review by the Editor-in-Chief of this journal.

Availability of data and materials

The data sets during the current study are available from the corresponding author on reasonable request.

Authors' contributions

AS, HE and MA: Collected clinical materials of these patients, participated in the design of the study and performed the statistical analysis. AG: Reviewed the analysis of echocardiographic and cardiac angiographic data. EE: participated in the study design, data analysis concerned with anatomy and pathophysiology and study coordination. All authors contributed to preparation of the clinical data used in this paper and revised the manuscript critically. All authors read and approved the final manuscript.

References

1. Moons P, Engelfriet P, Kaemmerer H, Meijboom F J, Oechslin E, et al. (2006) Expert Committee of EuroHeart Survey on Adult Congenital Heart Disease. Delivery of care for adult patients with congenital heart disease in Europe: results from the EuroHeart Survey. *Eur Heart J* 27: 1324-1330.
2. Hoffman JJ, Kaplan S, Liberthson RR (2004) Prevalence of congenital heart disease. *Am Heart J* 147: 425-439.
3. Macmahon B, Mckeown T, Record RG (1953) The incidence and life expectation of children with congenital heart disease. *Br Heart J* 15: 121-129.
4. Deanfield J, Thaulow E, Warnes C, Webb G, Kolbel F, et al. (2003) Task Force on the Management of Grown Up Congenital Heart Disease, European Society of Cardiology. ESC Committee for Practice Guidelines Management of grown up congenital heart disease. *Eur Heart J* 24: 1035-1084.
5. Webb GD (2001) Care of adults with congenital heart disease--a challenge for the new millennium. *Thorac Cardiovasc Surg* 49: 30-34.
6. Amaral FTV, Manso PH, Schmidt A, Sgarbieri RN, Vicente WVdeA, et al. (2015) Recommendations for starting a grown up congenital heart disease (GUCH) unit. *Bras Cir Cardiovasc* 30: 373-379.
7. Amaral F, Manso PH, Granzotti JA, Vicente WV, Schmidt A (2010) Congenital heart disease in adults: outpatient clinic profile at the Hospital das Clínicas of Ribeirão Preto. *Bras Cardiol J* 94: 707-713.
8. Berdat PA, Immer F, Pfammatter JP, Carrel T (2004) Reoperations in adults with congenital heart disease: analysis of early outcome. *Int J Cardiol* 93: 239-245.
9. Mello GA, Carvalho JL, Baucia JA, Filho JM (2012) Adults with congenital heart disease undergoing first surgery: prevalence and outcomes at a tertiary hospital. *Rev Bras Cir Cardiovasc* 27 : 529-534.
10. Monro J (2005) The changing state of surgery for adult congenital heart disease. *Heart* 91: 139-140.
11. Ghosh S, Chatterjee S, Black E, Firmin RK (2002) Surgical closure of atrial septal defects in adults: effect of age at operation on outcome. *Heart* 88: 485-487.
12. Lovell AT (2004) Anaesthetic implications of grown-up congenital heart disease. *Br J Anaesth* 93: 129-139.
13. Oliver JM, Gallego P, González AE, Benito F, Sanz E, et al. (2002) Surgical closure of atrial septal defect before or after the age of 25 years. Comparison with the natural history of unoperated patients. *Rev Esp Cardiol* 55: 953-961.
14. Jemielity M, Dyszkiewicz W, Paluszkiewicz L, Perek B, Buczkowski P, et al. (2001) Do patients over 40 years of age benefit from surgical closure of atrial septal defects? *Heart* 85: 300-303.
15. Webb G, Gatzoulis MA (2006) Atrial septal defects in the adult: recent progress and overview. *Circulation* 114: 1645-1653.
16. Amin Z (2006) Transcatheter closure of secundum atrial septal defects. *Catheter Cardiovasc Interv* 68: 778-787.
17. Amin Z, Danford DA, Pedra CA (2002) A new Amplatzer device to maintain patency of Fontan fenestrations and atrial septal defects. *Catheter Cardiovasc Interv* 57: 246-251.

18. Bruch L, Winkelmann A, Sontaag S (2008) Fenestrated occluders for treatment of ASD in elderly patients with pulmonary hypertension and/or right heart failure. *J Interv Cardiol* 21: 44-49.
19. Warnes CA, Williams RG, Bashore TM (2008) Atrial septal defect: ACC/AHA 2008 guidelines for the management of adults with congenital heart disease. *J Am Coll Cardiol* 52: 121.
20. Gianluca R, Giorgio R (2005) Congenital heart diseases in aged patients, clinical features, diagnosis and therapeutic indications based on the analysis of a twenty five year midline search. *Cardiology J* 13: 293.
21. Chang CH, Lin PJ, Chu JJ, Liu HP, Tsai FC, et al. (1998) Surgical closure of atrial septal defect. Minimally invasive cardiac surgery or median sternotomy? *Surg Endosc* 12: 820-824.
22. Piechowiak M, Piestrzeniewicz K, Goch J H (2008) Atrial arrhythmias—what is their effect on the clinical status and heart efficiency in adult patients after surgical correction of atrial septal defect type II? *Med Sci Tech* 49: RA87-92.
23. Simonneau G, Robbins IM, Beghetti M, Channick RN, Delcroix M, et al. (2009) Updated clinical classification of pulmonary hypertension. *J Am Coll Cardiol* 54: S43-54.
24. Diller GP, Gatzoulis MA (2007) Pulmonary vascular disease in adults with congenital heart disease. *Circulation* 115: 1039-1050.
25. Steele PM, Fuster V, Cohen M, Ritter DG, McGoon DC (1987) Isolated atrial septal defect with pulmonary vascular obstructive disease: long-term follow-up and prediction of outcome after surgical correction. *Circulation* 76: 1037-1042.
26. Sachweh JS, Daebritz SH, Hermanns B, Fausten B, Jockenhoewel S, et al. (2006) Hypertensive pulmonary vascular disease in adults with secundum or sinus venosus atrial septal defect. *Ann Thorac Surg* 81: 207-213.
27. Gatzoulis MA, Freeman MA, Siu SC, Webb GD, Harris L (1999) Atrial arrhythmia after surgical closure of atrial septal defects in adults. *N Engl J Med* 340: 839-846.
28. Kort HW, Balzer DT, Johnson MC (2001) Resolution of right heart enlargement after closure of secundum atrial septal defect with transcatheter technique. *J Am Coll Cardiol* 38: 1528-1532.
29. Attie F, Rosas M, Granados N, Zabal C, Buendía A, et al. (2001) Surgical treatment for secundum atrial septal defects in patients >40years old: A randomized clinical trial. *J Am Coll Cardiol* 38: 2035-2042.
30. Thilén U, Berling S, Varnauskas E (2000) Atrial septal defect in adults. Thirty-eight-year follow-up of a surgically and a conservatively managed group. *Scand Cardiovasc J* 34: 79-83.
31. Konstantinides S, Geibel A, Olschewski M, Görnandt L, Roskamm H, et al. (1995) A comparison of surgical and medical therapy for atrial septal defect in adults. *N Engl J Med* 333: 469-473.
32. Shaheen J, Alper L, Rosenmann D (2000) Effect of surgical repair of secundum-type atrial septal defect on right atrial, right ventricular, and left ventricular volumes in adults. *Am J Cardiol* 12:1395-1397.
33. Mikhail GW, Prasad SK, Li W, Rogers P (2004) Clinical and haemodynamic effects of sildenafil in pulmonary hypertension: acute and mid term effects. *Eur Heart J* 25: 431-436.