Natural history of atrial septal defect in the sixth decade : Study of 5 cases

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Histoire naturelle de la communication interauriculaire chez l'octogénaire : Etude de 5 cas

LA TUNISIE MEDICALE - 2013 ; Vol 91 (n°04) : 243-247

R É S U M É

Prérequis: Les communications interauriculaires (CIA) figurent parmi les cardiopathies congénitales les plus tolérées jusqu'à un âge adulte. Néanmoins, la découverte après 60 ans demeure exceptionnelle.
But: Notre étude descriptive s'est intéressée aux aspects cliniques et évolutifs des CIA de type ostium secundum (OS) chez l'adulte après la sixième décade.
Methodes: Nous avons procédé par une revue des dossiers des patients porteurs de CIA (OS) hospitalisés au service de cardiologie de l'hôpital La Rabta de Tunis entre les années 1985 et 2010, ensuite nous avons repéré parmi ces dossiers les cas de CIA (OS) découvertes après 60 ans. Une analyse descriptive des données cliniques et paracliniques a été effectuée chez ces patients avec le suivi des patients a comporté une évaluation dela qualité de vie par un questionnaire préétabli, un examen cardiovasculaire, une radiographie du thorax, un électrocardiogramme et une échocardiographie.
Resultas : Parmi 40 cas de CIA (OS) découvertes à un âge adulte référés à notre service, nous avons repéré 5 cas qui se sont manifesté après 60 ans. Il s'agit de 2 hommes et de 3 femmes. L'âge moyen dudébut des symptômes était de 62 ans. Les symptômes étaient représentés par une dyspnée d'effort associée à des palpitations dans 4cas et une douleur thoracique chez un seul patient. L'ECG révélait des signes d'hypertrophie ventriculaire droite dans tous les cas et une fibrillation auriculaire (FA) chez 2 patients. La radiographie du torax montrait une accentuation de la vascularisation pulmonaire périphérique dans tous les cas. L'échographie transthoracique couplée à la voie œsophagienne avait confirmé la présence d'un large defect avec un diamètre moyen de 20mm (10-32 mm). Une HTAP systolique sévére >60 mm Hg dans 2 cas, moyenne chez 2 patients (45-55 mmHg) et modérée dans un cas <45mmHg. Le rapport des débits pulmonaire et systémique (QP/QS) moyen était de 2,2. L'épreuve de contraste a détecté un shunt bidirectionnel dans un seul cas. L'étude hémodynamique par cathétérisme veineux réalisés dans chez tous les patients avait montré un enrichissement en oxygène dans l'OD et confirmé les autres données hémodynamiques non invasifs. Une fermeture de la CIA par voie chirurgicale a été effectuée chez 3 patients. Aucun décès périopératoire n'est survenu. Après un suivi moyen de 50±25 mois, nous déplorons un décès tardif survenu chez un patient qui était en stade d'insuffisance cardiaque globale en préopératoire. La plupart des survivants demeurent paucisymptomatiques avec un passage en FA dans 2 cas. Le contrôle échographique a éliminé un shunt résiduel chez les patients opérés, une stabilité des chiffres des pressions pulmonaires sauf chez un patient non opéré.
Conclusion: Après revue de lalittérature, nospatients s'avèrent parmi les survivants les plus âgés rapportés dans le monde. Une faible évidence persiste concernant les indications defermeture dela CIA après la sixième décade. Une approche rationnelle pour la prise en charge de la CIA chez l'adulte en particulier symptomatique nécessite une évaluation contrôlée du bénéfice tiré du traitement chez un tel groupe de patients ayant une comorbidité élevée.

Mots-clés

Communication inter auriculaire ; sujet âgé ; histoire naturelle

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S U M M A R Y

Aim: Atrial septal defect (ASD) is one of the most common causes of congenital heart disease manifested in adulthood.
Aim: To describe clinical and likelihood picture of adults over 60 years born with an ASD type II.
Methods: We performed a retrospective study of adult'sASD referred toourinstitutionfrom 1985 through 2010. Clinical, electrocardiographic, echocardiographic and hemodynamic data were reviewed. On follow up study, patients were investigated by echocardiography, ECG and assessed for quality of life by a questionnaire
Results: Among forty ASD type II manifested in adulthood which were referred to our department of cardiology five cases of ASD manifested in the sixth decades (2 men and 3 women). Complaints were dyspnea and palpitations in 4 cases and chest pain in only one patient. Slight anterior chest deformity was present in the older patient. Systolic murmur was found in the 3rd left intercostals space and the pulmonary second heart sound was accentuated in all patients. Complete right bundle branch block and right ventricular hypertrophy were found in all cases. Three patients presented atrial fibrillation. There was marked cardiomegaly in four patients. The pulmonary arteries were markedly enlarged and the peripheral vascular markings were increased. Echocardiographic data revealed large secundum ASD (mean 20 mm, ranged between 10 and 30mm), severe systolic pulmonary pressure in two cases (>5O mmhg). MeanQP/QS was 2.2 and contrast revealed bidirectional shunt in one patient. All patients were studied by venous cardiac catheterization. They showed a significant increase in the oxygen content of right atrial blood. Three patients underwent surgical atrial septal defect closure under general anesthesia. There were no operative or peri operative deaths. At mean follow up of 50±75 months, there was one late death from heart failure in a patient with advanced preoperative heart failure. The oldest patient is in the medical group and he is 75years old. Most survival patients remain in good clinical condition. Some of them were symptomatic at the last follow up and complained of shortness of breath on effort and palpitations in two cases. Two patients were in chronic atrial fibrillation developed during follow up. However, chest RX showed reduction in cardiothoracic ratio postoperatively. Echocardiographic examination confirmed that there was no residual shunt in across the atrial septum in any patient. Systolic pulmonary pressure felled only in 2 patients in the surgery group.
Conclusion: To our knowledge, thesepatients havealongue life span, although survivors with ASD described in the world. There is a lack of evidence regarding treatment options for adults with an ASD aged more than 60years. Given the higher risks of surgery in advanced age, the defect should be repaired as early as possible to prevent hemodynamic complications.

Key-words

Atrial septal defect; adulthood; natural history

Atrial Septal defect (ASD) is one of the most common causes of congenital heart disease manifested in adulthood. Among the three kinds of ASD, the secundumtype ASD account for two-thirds of all such defects (sinus venosis, ostium primum, ostium secundum and unroofed coronary sinus ASD type). The defect is compatible with a normal life span-survival up to years (1-3) and natural history of ASD over the age sixteen remains unclear because incomplete information regarding of. Closure of most atrial septal defects is still the treatment of choice in children and young adults. However, the beneficial resultof closure in adults over 60 years of age remains controversial.

We carried out the present study to discuss natural history in this group of patients on the light of few cases reports and review of the literature.

METHODS

We performed a retrospective analysis of 40 adultswith born ASD admitted from 1985 through 2010 at institution of adult cardiology in Rabta Hospital. We studied only patients with ASD and without any other cardiac abnormalities. Partial anomalous pulmonary venous drainage was not regarded as a complication.Clinical, electrocardiographic, chest radiographic, echocardiographic and Hemodynamic data were reviewed.In The follow up, Patients were investigated by echocardiography, ECG and assessed for quality of life by a pre established questionnaire and a complete physical examination.

The search engines which have been used were: Pub-Med, Cochrane, and Scopus. We have limited the research to the last twenty years.

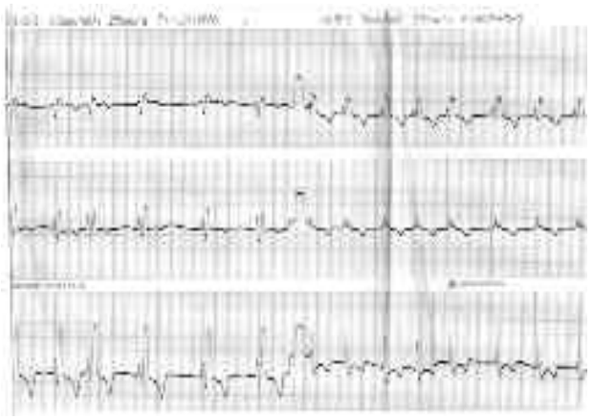
RESULTS

Among forty patients with ASD manifested in adulthood, we have individualized five octogerian cases. Table I shows the clinical and haemodynamic data of the patients.The majority of them were female. Age ranged between 60 and 75 years. The age at onset of symptoms varied from sixty-for to seventy- five years.

Clinical features at presentation

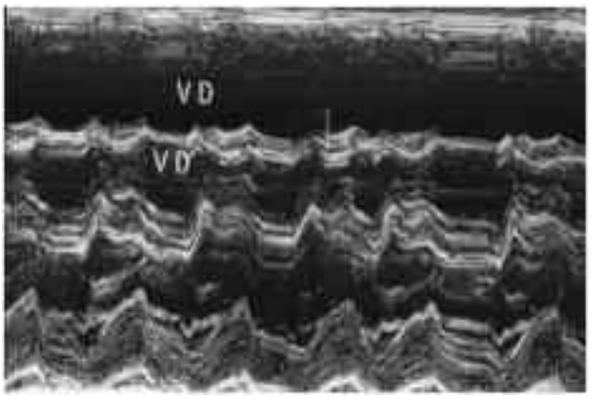
The presenting symptoms were dyspnea in all patients and palpitations in two. Chest pain occurred in only one patient.Slight anterior chest deformity was present in the older patient. Systolic murmur was found in the 3thd left inter costal space and the pulmonary second heart sound was accentuated in all patients. The neck veins were distended, the liver was enlarged and Harzer sign was found in one case. Electrocardiographic patterns were presented by right ventricular hypertrophy as characterized by right deviation of electric axis of the heart, complete right bundle branch block andwere found in all cases.Two patients presented atrial fibrillation at presentation (figure1). Chest radiography showed marked cardiomegaly in two patients (cardiothoracic ratio> 0.65) and moderate in the other cases (<0.6). The pulmonary arteries were markedly enlarged, the peripheral vascular markings were increased and right atrial and right ventricular enlargement were noted in all cases in the absence of significant left atrial enlargement.

Figure 1 : ECG: Complete right bundle branch block



Transthoracic echocardiography (TTE) combined to transoesophageal echocardiography (TOE) confirmed the presence of secundum ASD. Medium atrial septal defect diameter was 20 mm ranged between7 and 31mm (Figures 2 and 3).

Figure 2 : Tranthoracic echography mode TM : - Right ventricular dilatation - Paradoxical septal wall motion



Right ventricular dilatation and paradoxical septal wall motion were found in all patients. The pulmonary artery was markedly dilated (Figure 3). The ratio of pulmonary/systemic output was 2.7 ± 0.6 (range 1.7 to 7.4). Pulmonary systolic pressure was 32.3 ± 15 mmHg. Associated anomalies were mitral valve prolapse in one patient, aortic rheumatic valve deformities in two patients and anomalous pulmonary venous return in one case.TOE with contrast revealed bidirectional shunt in one case. Two patients studied by venous cardiac catheterization showed an increase in the oxygen content of right atrial blood. Catheter was successfully passed a cross an ASD. Pulmonary hypertension was moderate (figure 5) and severe only in two cases. Small right to left shunt was found in one case.

Figure 3 : Transoesophageal echocardiography: large ASD ostium secundum

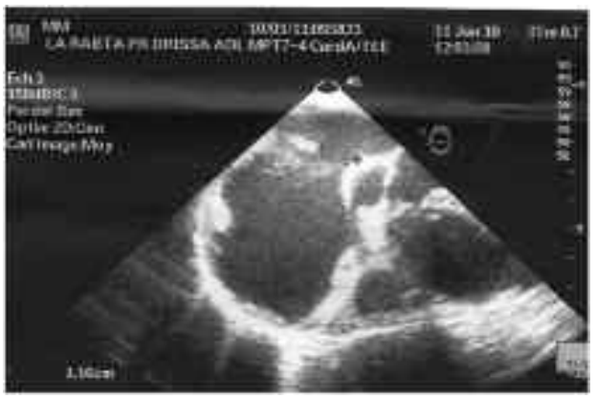
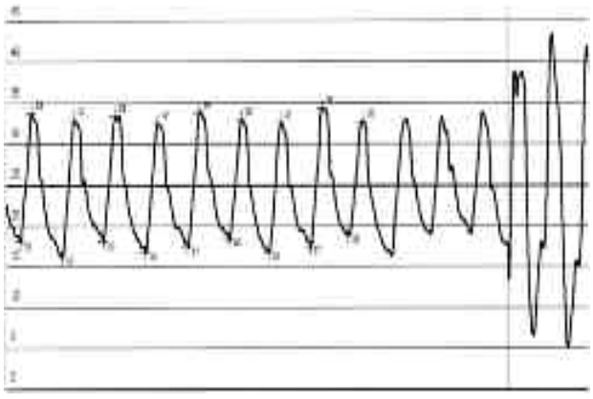


Figure 4: Moderate systolic pulmonary arterialhypertension showed on manometric curve in catheterism laboratory



Outcome of patients

There were no operative or perioperative deaths. At mean follow up of 50 ± 25 months, there was one late death from heart failure in a patient with advanced preoperative heart failure. Among the survivors, the oldest patient in the medical group is 73 yearsand in the surgical group is 85 years old. Most survival patients remain in good clinical condition. Some of them were symptomatic at the last follow up and complained of shortness of breath on effort and palpitations in two cases. Patients were in chronic atrial fibrillation developed during follow up. However, chest RX showed reduction in cardiothoracic ratio postoperatively.Echocardiographic examination confirmed that there was no residual shunting across the atrial septum in any patient.

DISCUSSION

After bicuspid aortic valve, ASD is themost common congenital defect in adults. It was first described by Rokitansky in 1875 cited by Edwards et al (4) and became well known with the writings of Barber et al (5) and Wood (6) in the 1950s. Fuster et al (7) reported an incidence of 22% among adults with congenital heart disease, but the true incidence is unknown as it isoften not recognized until late in life.But, with the increased detection rate of congenital heart disease in adults,which has resulted from more medical examinations and the availability of echocardiography, the real incidence is certainly higher.

Clinical picture of ASD in adult is different compared to young patients.In elderly patients, associated diseases, such as coronary heart disease, arrhythmias and pulmonary arterial hypertension were present in some cases. Differentiation between complications of ASD and heart geriatric diseases is so difficult in routine practice .Symptomatic deterioration has been attributed to the size of the defect (8), pulmonary hypertension, increasing pulmonary vascular resistance (9), atrial fibrillation, systemic hypertension, recurrent infection, and reduced left ventricular compliance (10, 11). Perloff (3) concluded that a combination of reduced left ventricular compliance, atrial fibrillation, and mild to moderate pulmonary hypertension associated with a continuing large shunt is responsible of heart failure onset.

In1970 Campbell re analyzed and extended the data (10). He calculated the annual mortality to be approximately 0.6% in the first and second decades, 6.3% in the fourth decade, and 9.4 % in the sixth and subsequent decades. The next published report concentrated on the older age groups with a study of 67 patients who survived over 40. Pulmonary hypertension was rare in young patients but increased in frequency with age. 30% of patients in the sixth decades had a pulmonary artery systolic pressure of > 50 mmHg. Approximately 40 % of patients had died or were seriously disabled in the fifth decade and 90% of survivors aged over 60 years were severely limited. Deterioration was attributed to atrial fibrillation, recurrent bronchitis and pulmonary infarction.

The natural history reports by Craig and Selzair (12) is the most detailed and provides haemodynamic and clinical data on128 patients aged 18-56. The quarters of patients had symptoms. Most were dyspnoeic. Pulmonary artery systolic pressure was greater than 25 mmHg in 60% of cases but only 15% had obstructive pulmonary hypertension (pulmonary vascular resistance> 5 units. The authors concluded that pulmonary hypertension was the commonest single factor to presage worsening symptoms.

Significant reductions in the pulmonary arterial pressure have been observed in the post-operative hemodynamic studies reported (13, 14).Interestingly the reduction occurred in all age groups and functional classes, but was more significant in patients with moderate or severe pulmonary hypertension.

In a multivariable model of an epidemiological study of a large European cohort of ASD type II in adulthood (14), the best independent predictors of functional limitations appeared to be pulmonary arterial hypertension (PAH) with an odds ratio 25.2 (5.8-109.6); P<0.001) and right ventricular volume overload with an odds ratio 2.3 (1.5-3.4; P<0.001) . Moderate to severe hypertension was found in contrast to usual finding of normal pulmonary artery pressure in the young age. In our study, PAH was found in all cases which was severe in cases. Our oldest

patient, aged 76 years, had markedly increased systolic PAP (> 100 mmHg).

The question that remains is whether or not there is a haemodynamic benefit from closure of the atrial septal defect.

Kannel and al (15) have noted the frequent combination of atrial fibrillation and cardiac failure with increasing age. Comparison between surgical and medical groups according to atrial fibrillation, embolic episodes, and cardiac failure provided information on possible reduction of complications of ASD by surgical closure. However, in a previous study (16) of patients with ASD aged >= 40 years, authors showed that overall mortality was not different between operated and not operated patients, although there was a tendency in favor of operated patients. However, Morbidity was higher in not operated patients. Surgical closure is therefore, feasible in this age group. Significance association of cardiac and pulmonary disease, however must be considered.

In a historical, prospective study (14), of first diagnosed ASD after the age of 25,outcome in adultsafter medical or surgical treatment was compared by Shah et al. There was no difference in survival or symptoms between the two groups and no difference in the incidence of new arrhythmias, stroke or other embolic phenomena, or cardiac failure. No patient in either group developed progressive pulmonary vascular disease. For Shah, progressive pulmonary vascular disease did not develop in any of these patients. Its prevention is not a reason for advising closure of ASD in adults.

For Landi (17), taken together, non-operated patients fared significantly worse in all aspects of hemodynamic studied than the patients whose defects had been closed. Despite the frequency of complications, it has been concluded that surgically treated patients fare better than those treated medically. The evidence on which this conclusion is based does not stand up to close scrutiny.

Steel (18) found that after a minimum follow up of four years 20 % of surgically treated patients and 71% of medical patients had dad. Not surprisingly they concluded that, except for those in whom pulmonary vascular resistance was more than 10 units, surgery was the correct treatment.

In the study reported by Wolf et al (19) ,20% of the surgicalgroup died post operatively, 25% died later, but 50% were improved. After an unstated time, 68% of traceable medical cases were alive, nine of whom had minimal or no symptoms. In both of these studies patients in the medical group were more seriously ill. Despite deficiencies in the data, their conclusion, that surgical mortality in patients with severe pulmonary hypertension is high, is widely accepted. Their other conclusion, that symptom free adults may fare better with medical treatment, finds support from Shah et al (14) who compared similar groups of medically and surgically treated adults followed for 20 years Surgical patients fared no better over the 20 years of follow up than medical cases with respect to mortality and to the incidence of breathlessness, atrial fibrillation, emboli, and cardiac failure.

The patient with a bidirectional shunt will usually tolerate theclosure, although postoperative pulmonary complications and alonger period of morbidity are morelikely to occur. The use of pulmonary vasodilators in the postoperative period in such patients and vigorous postoperative chest physiotherapy are usually helpful. The rare patient who is visibly cyanotic and he is a rare patient indeed has irreversible pulmonary vascular changes andwill not tolerate closure. The age beyond which surgery should not be routinely offered is uncertain because the situation in adults is less clear than in children. A rational approach to the management of atrial septal defect in adults, in particular those with symptoms, requires a controlled assessment of the relative merits of medical and surgical treatment.

CONCLUSION

There is a lack of evidence regarding treatment options for adults with an ASD aged more than 60 years.Surgical correction of ASD is associated with low morbidity and mortality. However, given the higher risks of surgical closure of the defect in advanced age, the defect should be repaired as early as possible to prevent hemodynamic complications, such as the development of pulmonary hypertension and cardiac arrhythmias. Surgical intervention in people with few or no symptoms must be proved to be of benefit. Such benefit has never been shown in a randomized prospective series of adults with ASD.

Références

1. Seldon WA, RubsteinC,Fraser AA. The incidence of atrial septal defect in adults. Br Heart 1962; 24:552-62.
2. Colmers RA. Atrial septum defect in elderly patients: report of three patients aged 68, 72, and 78 years. Am J Cardiol 1958;1:768-73.
3. Perloff JK. Ostium secundum atrial defect-survival for 87 and 94 years. Am J Cardiol 1984;53:388-9.
4. Edwards JE, Carey LS, Neufeld HN, et al. Congenital heartdisease: correlation of pathologic anatomy and angiograph.Philadelphia WB Saunders, 1965,1:191-234.
5. Barber JM, Magidson D, Wood P. Atrial septal defect. BrHeartJ 1950;12:277-9.
6. Wood P. Diseases of the heart and circulation ed.2, p 358 London 1956.
7. Fuster V, Brendenburg RO, McGoon DC, et al. Clinicalapproach and management of congenital heart diseasein the adolescent and adults. Cardivasc Clin 1980; 10: 161-97.
8. 8/ Bedford DE, Papp C, Parkinson J. Atrial septal defect. Pulmonary hypertension increasing pulmonary vascular resistance Br Hearty 1941;3:37-67
9. Dexter L. Atrial septal defect. Br HeartJ 1956;18: 209-25.
10. Campbell M, Neill C, Suzman S. The prognosis of atrial septal defect. Br Med J 1957;i: 1375-83
11. Konstantinides S, Geibel A, Kasper W, et al. The natural course of atrial septal defect in adults-a still unsettled issue. Kin Wochenschr 1991; 69:506-10.
12. Craig RJ, Selzer A. Natural history and prognosis of atrial septal defect. Circulation 1968;37:805-15.

13. Decoff GR, Brandenburg RO, et al. Results of operation for atrial septal defect in patients forty-five years and older. Circulation1967; 35:143-7.
14. 14/ Murphy JG, Gersh BJ, Mc Goon MD, et al. Long term outcome after surgical repair of isolated atrial septal defect. N Engl J Med 1990; 323:1644-50.
15. 15/ Kannel WB, Aboot RD, Savage DD,MC Namara AF. Epidemiologic features of chronic atrial fibrillation. N Eng J Med 1982;306:1018-20.
16. 16/ Shah D, Azhar M, Oakley CM, et al. The natural history of medically and surgically treated adults with secundum atrial septal defect. A historical prospective study. Br Heart J 1994;71:224-8.
17. 17/Landi F, Cipriani L, Cocchi A, Zuccala G, Carbonin P. Ostium secundum atrial septal defect in the elderly. J Am Geriatr Soc. 1991;1:60-3.
18. 18/ Steel PM, Fuster V, Cohen M, et al. Isolated atrial septal defect with pulmonary vascular obstructive disease. Long term follow up and prediction of surgical outcome after surgical correction. Circulation 1987;76:1037-42.