Large Atrial Septal Defects in Adolescents and Adults: A High Incidence of Sinus Venosus Type Defects Presenting Late

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Background: Isolated Atrial Septal Defect (ASD) is a common congenital heart disease that could present at any age. It is the most common form of heart disease presenting in adolescents and adults.

Objective: To evaluate adolescents and adults presenting late with significant ASD's and their presentation, management and short-term outcomes.

Design: A Retrospective Study.

Setting: Mohammed Bin Khalifa bin Salman Al-Khalifa Cardiac Center, Bahrain.

Method: A retrospective study was performed. Adolescents/adults more than 14 years of age with significant ASD referred to a tertiary cardiology unit over a three-year period were reviewed from 1 March 2012 to 31 March 2015. The data was obtained from a computerized database.

Result: Thirty-two patients presented with large ASD. Twenty-five (78.1%) were females. The mean age was 29.4 years. The majority were diagnosed as an incidental finding of murmur during routine examination. A high number of Sinus Venosus Defects in the adolescent/adult group (22% compared to 7% in the pediatric group: P=0.01) was found, which were more common in males (42% compared to 17%: P=0.128). Nine (28.1%) underwent successful device closure. Twenty-three (71%) patients were referred for surgical correction. There was no mortality.

Conclusion: Significant ASD is often undiagnosed until adolescence or adulthood. If not promptly diagnosed and managed, it is likely to present with symptoms and signs of pulmonary overload later in life. Sinus Venosus ASD's should be suspected in male adolescents and adults with significant ASD's in Bahrain and surgical closure is often required.

Bahrain Med Bull 2017; 39(1):17 - 19

Isolated Atrial Septal Defect (ASD) is a common congenital heart disease and could present at any age. It represents 7% of all cardiac defects and is the most common form of heart disease presenting initially in adolescents and adults[1,2].

Large ASD's with right heart dilatation left unclosed have an important age-related morbidity and mortality. Recent advances in the diagnosis and management of isolated ASD's have benefitted these patients greatly. This study reviews the late presentation of adolescents and adults in the Kingdom of Bahrain.

The aim of this study is to evaluate adolescents and adults presenting late with significant ASD and their presentation, management and short-term outcomes.

METHOD

Patients over 14-years of age with significant ASD referred to a tertiary cardiology unit over a three-year period were reviewed from 1 March 2012 to 31 March 2015. The data was obtained from a computerized database. All relevant data was analyzed.

Statistical analysis was performed using SPSS version 6. The grouped data were compared using Chi-square test. The null hypothesis was rejected when P-value>0.05.

RESULT

One thousand two hundred twenty-two patients were referred to the specialist congenital cardiology service during the 3-year

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study period. One hundred forty patients had significant ASD; 32 were over 14 years of age presenting for the first time. Thirty-two adolescents and adults are the subjects of this study; 25 (78.1%) were females.

The age ranged between 14 to 54 years and the mean age was 29.4 years. The majority were Bahraini nationals, see figures 1, 2 and 3.

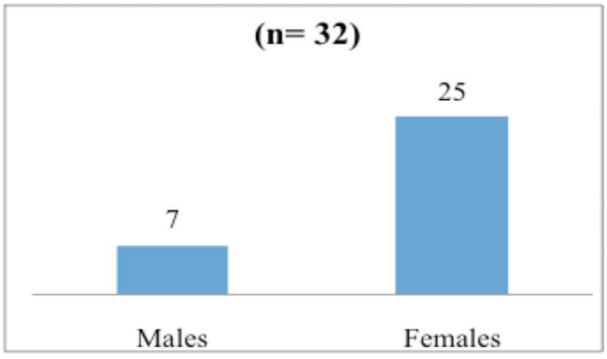


Figure 1: Gender Distribution

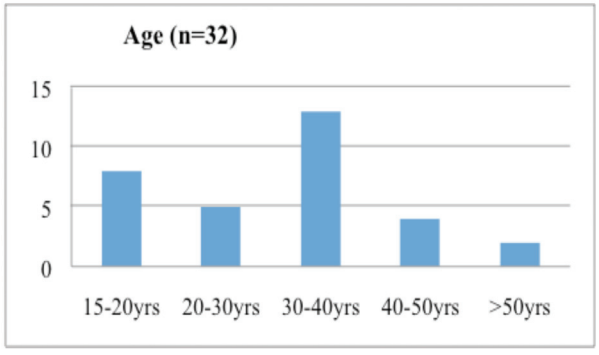


Figure 2: Age Distribution

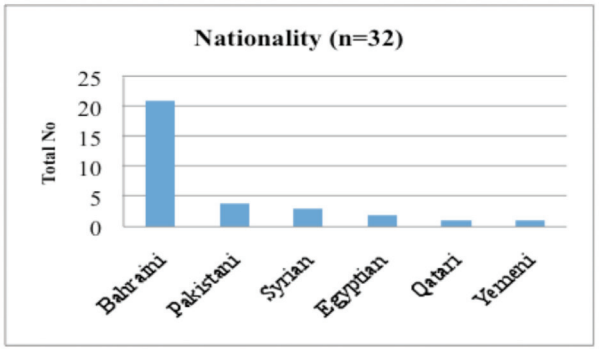


Figure 3: Nationality

The majority of the patients were discovered as incidental finding of murmur during routine examination 14 (43.7%), palpitations 8 (25%), dyspnea on exertion 8 (25%), cerebral vascular accident 1 (3.1%) and brain abscess 1 (3.1%). No patients were on any medications prior to presentation.

Patients underwent full clinical evaluation, ECG, CXR and transthoracic evaluation, which confirmed the diagnosis of a large significant ASD.

The majority of the defects were ASD of the secundum, but there was a high number of Sinus Venosus type defects. The study confirmed a high number of Sinus Venosus Defects in adolescent and adult patients compared to pediatric patients (22% compared to 7 %, P=0.01). Furthermore, the Sinus Venosus Defects were more common in males (42% compared to 17%, P=0.128), see figures 4, 5 and 6.

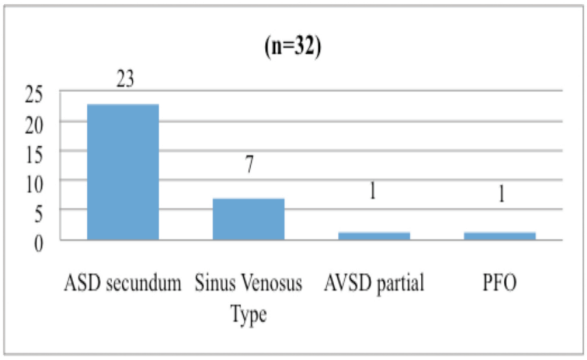
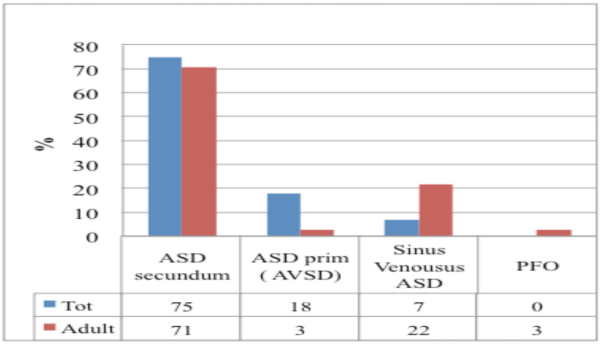
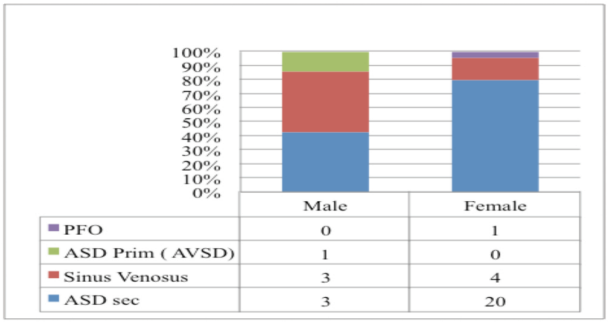


Figure 4: Types of ASD



ASD sec = Atrial Septal Defect Secundum Type
ASD prim (AVSD) = Primum Atrial Septal Defect
PFO = Patent Foramen Ovale

Figure 5: Total and Adult Type of ASD



PFO = Patent Foramen Ovale
Prim ASD = Primum Atrial Septal Defect
AVSD = Atrioventricular Septal Defect
ASD sec = Atrial Septal Defect Secundum Type

Figure 6: Types of ASD and Gender

Evaluation with Trans Esophageal Echocardiogram (TOE) was performed to delineate the anatomy and/or assess the viability for possible trans-catheter delivered device closure.

Thirteen (40.6%) patients had cardiac catheterization to attempt device delivered closure. Nine (28.1%) had successful device closure with a variety of Amplatzer Septal Occluder device sizes, see figure 7. One (3.1%) patient failed device implantation with embolization to the right ventricle that required surgical removal and ASD closure. Three (9.4%) patients were found unsuitable and no attempt at closure was made; therefore, they were referred for surgical closure.

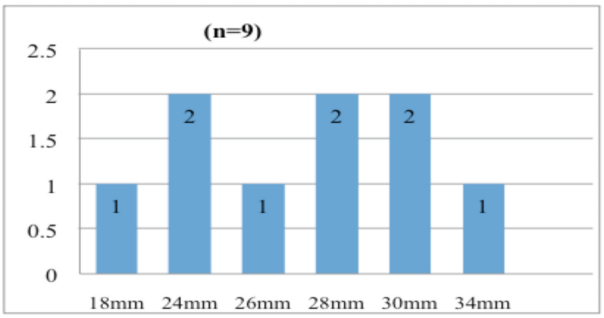


Figure 7: Device Size

Twenty-three (71.9%) patients were referred for surgical correction, see figure 8. Seven (21.9%) Sinus Venosus and one (3.1%) partial AVSD type ASD were referred primarily for surgery. Fifteen (46.9%) secundum type ASD were also referred for surgery because 11 (34.4%) were found to be unsuitable for device closure after initial TOE evaluation found either to have inadequate ASD rim anatomy or too large ASD size. One (3.1%) patient had failed device closure and the three (9.4%) were planned for catheter-delivered device, but were found to be unsuitable during cardiac catheterization were also referred for surgical closure. No mortality was recorded with device closure or surgery.

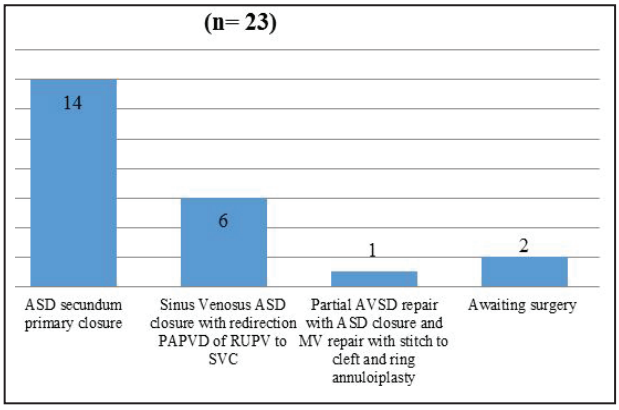


Figure 8: Surgical Closure

DISCUSSION

Three major types of interatrial defects could be found: Ostium primum defect, Ostium Secundum defect and Sinus Venosus defect. Ostium Secundum defects are the most commonly reported, occurring in about 1/1500 livebirths[3]. The Sinus Venosus ASD normally constitutes only 5% to 10% of all ASD's[4].

The reason for the high number of Sinus Venosus ASD noted in our study (22%) is unclear. It appears not to be related to demographic reasons alone as the overall number (pediatrics) of these types of defects during the study periods was 7%, which is similar to previous studies[5]. Diagnosis of Sinus Venosus ASD is often difficult compared to other types of ASD's and should be suspected in any adolescent/adult with unexplained right heart dilatation. Cardiac MRI is occasionally needed to confirm the diagnosis if the Transesophageal Echocardiogram is not confirmatory; that was not required for any of our patients.

Currently, the only treatment for Sinus Venosus ADS's is surgical closure due to the common connection of some or all of the right sided pulmonary veins to the superior vena cava or right atrium that needs to be redirected. This accounts primarily to the high number of patients in this study (72%) requiring surgical closure of their ASD's rather than catheter delivered device (28%) compared to other series.

CONCLUSION

Significant ASD's often go undiagnosed until adolescence or adulthood. These adults with isolated ASD's commonly reach adulthood before being diagnosed. They are becoming a relatively large population. If they are not correctly diagnosed and managed, they are likely to present with symptoms and signs of pulmonary overload later in life. Due to the high number of male adolescents and adults with Sinus Venosus ASD's in Bahrain, surgical closure is more often required rather than catheter delivered device closure.

Author Contribution: All authors share equal contribution towards: (1) substantial contributions to conception and design, acquisition of data, or analysis and interpretation of data; (2) drafting the article or revising it critically for important intellectual content; and (3) final approval of the version to be published. Yes.

Potential Conflicts of Interest: None. **Competing Interest:** None.

Sponsorship: None.

Acceptance Date: 6 February 2017.

Ethical Approval: Approved by the Mohammed Bin Khalifa bin Salman Al-Khalifa Cardiac Center, Bahrain Defense Force Hospital, Bahrain.

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