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Echocardiographic Morphological Classification of Aortic Stenosis in Sulaimani Pediatric Teaching Hospital/Kurdistan/Iraq

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Abstract: ***Aortic stenosis occurs when the heart's aortic valve narrows. This narrowing prevents the valve from opening fully, which obstructs blood flow from the heart into the aorta and onward to the rest of the body. The aim of the study was to performed the echocardiographic study of the major morphological types of aortic valve stenosis in the pediatric cardiac unit in suliamani. A retrospective study including 127 patients aged from birth to 14 years done in the cardiac unit / Sulaimani pediatric teaching hospital. The data were collected from the recorded files of the patients examined in that unit during the period from 2006 to 2016.Collected data included name, age, sex, residency, consanguinity, clinical presentation, associated syndromes and Echo findings. Statistical analysis was performed by SPSS 21. Chi-square test was used to find out the correlation between categorical variables, P value of (≤ 0.05) was regarded significant. Overall, 80 were males (63%) and 47 were females (37%). Forty patients (31.5%) were below one year. The valvular type found in 96 cases (75.6%). subvalvular and supravalvular types found in 22 cases (17.3%), 9 cases (7.1%) respectively. We found 91 cases (94.8%) of bicuspid, 3 cases (3.1) of tricuspid and 2 cases (2.1%) of the unicuspid valve. In subvalvular type we found 14 cases (63.6%) of sub aortic ridge, 5 cases (22.7%) of tunnel type and 3 cases (13.6%) of systolic anterior motion. In supravalvular type we had 7 cases (77.8%) of hourglass and 2 cases (22.2%) of the long segment. Aortic regurgitation was the most common associated cardiac defect. There was a significant association between the types of aortic stenosis and the mild grade of AR; P value <0.05. In conclusion the valvular aortic stenosis was the most common type of aortic stenosis in this study. Bicuspid aortic valve found to be the most common congenital anomaly associated with aortic stenosis. Most of the patients with aortic stenosis were discovered to have an accidental murmur.***

Keywords: Stenosis, Echocardiography, Bicuspid, Valvular, Regurgitation, Congenital.

1. INTRODUCTION

Congenital aortic stenosis occurs in different forms, usually classified with respect to the site of the obstruction relative to the aortic valve: valvular, subvalvular and supravalvular [1].

Congenital aortic valvular stenosis itself forms 5% of all cardiac abnormalities. It is difficult to know the true incidence, however, because of congenitally abnormal aortic valves may not be recognizedinchildhood [2].

It is more common in males with male to female ratio of 4:1. Associated anomalies can be detected in as high as 20% of patients [3].

Echocdiography defines the anatomy of the aortic valve and severity of aortic valve obstruction. Left ventricular muscle is hypertrophied depending on the severity of aortic valve obstruction. Doppler flow velocity across the aortic valve is useful tool in assessing the severity and planning the intervention [3].

Subvalvular aortic stenosis:

Fixed subvalvular aortic stenosis accounts for approximately 10% to 20% of patient of aortic stenosis in children, and the male to female ratio is 2:1 to 3:1. Associated cardiac defects are present in more than half of patients. Common associated cardiac defects include VSD, coarctation of the aorta, atrioventricular septal defect**, and valvular aortic stenosis.**

Most commonly, a discrete fibrous membrane or fibromascular collar encircles the left ventricular outflow tract just beneath the aortic valve . Rarely, a long diffuse fibro muscular obstruction narrows the left ventricular outflow tract for several centimeters, forming a tunnel sub aortic stenosis [4].

*Echocardiography*with Doppler study is highly sensitive and specific for the diagnosis of sub aortic stenosis and to define the anatomy of the lesion Diagnostic cardiac *catheterization*is not mandatory in the evaluation of sub aortic stenosis, but may provide useful information in cases where the clinical data and noninvasive evaluation are not consistent [1].

Supravalvular aoertic stenosis:

Supravalvular aortic stenosis is the least common type of aortic stenosis. It is rare. Approximately 30% to 50% of patients with supravalvular aortic stenosis have Williams- Beuren syndrome [1].

The anatomic diagnosis of SVAS can reliably be made from two- dimensional echocardiography that uses multiple views, including parasternal, apical long- axis, and suprasternal. In SVAS with hourglass deformity and diffuse hypoplasia, the diameter of the ascending aorta is smaller than that of the aortic root. In SVAS with fibrous diaphragm, the external ascending aortic diameter is

normal, although an echogenic membrane is commonly observed above the sinuses of Valsalva. Turbulent color flow mapping indicates the site of hemodynamically significant obstruction in relation to the origin of the coronary Ostia. The incidence of coronary artery involvement is high in SVAS [5]. Doppler peak gradient overestimates and, therefore, does not predict Catheter-measured gradient well in patients with SVAS and may not be dependable in assessing its severity and guiding the need for intervention [6].

2. METHODS AND MATERIALS

This is a retrospective study including 127 patients aged from birth to 14 years; diagnosed as cases of aortic valve stenosis: conducted in the pediatric cardiac unit / sulaimani paediatric teaching hospital.Thedata werecollected from the recorded files of the patientsexaminedin that unit during 10 yearsfrom 2006 to 2016.

The data of study included name, age, sex, residency, and consanguinity, symptoms of the presentation which include (Accidental murmur, fatigability, chest pain, syncope, shock and feature of congestive heart failure). Associated syndromes included William Syndrome (Elfin facies, intellectual disability,heart problems, especially supravalvular aortic stenosis, and periods of high blood calcium),Turner Syndrome(Webbed neck, low-set ears, low posterior hairline, shield chest ,short stature, and lymph edema), Crouzon

A_ Acquired: SAM

Sub aortic tissue ridge

B _Congenital (tunnel type)

supravalvular aortic steno

All cases included in this study were examined by the same ECHO machine using Two dimensional, and Doppler(spectral and color) with 3V2C and 7V3C MHz transducers (adjusted according to examinations were obtained for each patient chest wall thickness) using a commercial Accuson Cypress, USA made, supplied by Siemens Company and by the same examiner (pediatric cardiologist). Trans-thoracic cross-sectional echocardiography usually allows clear demonstration of all the intra cardiac anatomy. Echo done according to standard echo views from sub costal, apical, parasternal, suprasternal position according to the standards recommended by the American Society of Echocardiography [7]. All patient's data entered using computerized statistical software; statistical package for Social Science (SPSS) version 21 for windows. Chi - square test was used for categorical variables, p value of $\leq .005$ was regarded as significant and the results presented as tables.

3. RESULTS

We have 127 cases of aortic valve stenosis, 80 patients were males (63.0%), while 47 patients were females (37.0%). The ages ranged from birth to 14 years, 40 pts. wereless than one year (31%).As shown below,88 patients (69.3 %) were from sulaimani. All these details mentioned in table (1).

Table 1. Descriptive features regarding sex, age and residency

VARIABLES	NUMBERS	%
SEX		
Male	80	63.0
Female	47	37.0
Total	127	100.0
AGE		
Less than one year	40	31.5
1-5 years	30	23.6
6-10 years	30	23.6
10-14years	27	21.3
Total	127	100.0
RESIDENCE		
Sulaimani	88	69.3
Out of sulaimani	39	30.7
Total	127	100.0

Table (2) shows that **96** cases(75.6%) were VAS, which makes the most common type of AS, while **22** cases(17.3%) were SAS and **9** cases (7.1%) were SVAS. BAV cases were found in **91** patients (94.8 %); the most common type of VAS. The cases of SATR were **14** (63.6%), makes it the most common type of SAS. In pts with SVAS, cases of hourglass were **7** (77.8 %); the most common one.

Table 2. Descriptive features regarding the morphological classification of AS.

Variables	NO.	%
VAS	96	75.6
SAS	22	17.3
SVAS	9	7.1
Total	127	100.0
VALVULAR AORTIC STENOSIS		
Unicuspid	2	**2.1**
Bicuspid	91	94.8
Tricuspid	3	3.1
Total	96	100.0
SUBVALVULAR AORTIC STENOSIS		
SAM	3	13.6
Sub aortic tissue ridge	14	63.6
Congenital tunnel type	5	22.7
Total	22	100.0
SUPRAVALVULAR AORTIC STENOSIS		
Hourglass	7	77.8
Long segment	2	22.2
Total	9	100.0

Table (3) shows that 57 cases (44.9%) of the patients presented with accidental murmur. Other clinical features were included CHF in 33 cases (26%), fatigability which is seen in **19** cases (15%), chest pain in 11 cases (8.7%), critical AS with shock in 4 cases (3 %) and Syncope in 3 cases (2.4%).

Table 3. patterns of clinical presentations

variable	***Number***	***%***
Accidental murmur(auscultation)	57	44.9
CHF	33	26
Fatigability	19	15
Chest pain	11	8.7
Shock	4	3
Syncope	3	2.4
Total	127	100

There is a significant association (p **value < 0.05**) between the cases presented with features of CHF and the types of aortic valve stenosis; as 32 cases of VAS presented with congestive heart failure, as shown in table (4).

We have **7** cases of Williams's Syndrome, **3** cases with Turner Syndrome, **2** cases of Down Syndrome and one case of Crouzon disease. There was a significant association between Williams's Syndrome and the types AS (**p value < 0.05**). There was no significant association between the types of aortic stenosis and other syndromes (P value >0.05),as mentioned in table(5).

The most common associated cardiac defect in patients with valvular and subvalvular aortic stenosis was aortic regurgitation; while we had no reported cases of SVAS associated with AR; in spite of that VSD and PDA were the most common associated cardiac anomalies in those patients (SVAS). Cardiac anomalies were reported in all cases of SAS (No isolated cases).These details and other associated cardiac defects arranged in table (7)

We found that **49** cases of aortic valve stenosis associated with aortic regurgitation; which further subdivided according to the severity into mild, moderate and sever AR. The mild one was the most frequently encountered defect; which is detected in **38** cases of VAS and SAS. There was a significant association between the types of AS and the mild grade of AR **(p value < 0.05**). Table (8) does clarify all these data.

Table 4. Association between the types of AS and patterns of clinical presentation.

Variable		VAS	SAS	SVAS	P value
Accidental murmur	Yes	38	13	6	0.100
	No	54	9	3	
Fatigability	Yes	11	6	2	0.141
	No	85	16	7	
CHF	Yes	32	1	0	0.004
	No	64	21	9	
Chest pain	Yes	10	1	0	0.428
	No	79	18	8	
Syncope	Yes	1	1	1	0.124
	No	95	21	8	
Shock	Yes	4	0	0	0.513
	No	92	22	9	

Table 5. Association between associated syndromes and types of aortic stenosis

Variable		VAS	SAS	SVAS	P value
Turner Syndrome	YES	3	0	0	0.609
	No	93	22	9	
Williams Syndrome	YES	0	0	7	0.000
	No	96	22	2	
Down Syndrome	YES	2	0	0	0.720
	NO	94	22	7	
Crouzon Syndrome	YES	1	0	0	0.850
	NO	95	22	9	

Table 6. Association between sex and consanguinity with the types of aortic stenosis.

VARIABLES		VAS	SAS	SVAS	P VALUE
Sex	Male	59	14	7	0.624
	Female	37	8	2	
Consanguinity	Positive	47	9	3	0.566
	Negative	49	13	6	

Table 7. Associated echocardiographic cardiac defects.

VARIABLE		Types of AS		
		VAS	SAS	SVAS
AR	*NO*	34	15	0
	%	34.4	68.2	0
COA	*NO*	11	2	0
	%	11.5	9.1	0
MR	*NO*	5	3	0
	%	5.2	13.6	0
PDA	*NO*	4	0	2
	%	4.2	0	22.2
VSD	*NO*	4	2	2
	%	4.2	9.1	22.2
HLHS	*NO*	2	0	0
	%	2.1	0	0
DCM	*NO*	2	0	0
	%	2.1	0	0
ASD	*NO*	1	0	0
	%	1	0	0
TR	*NO*	1	0	0
	%	1	0	0
PS	*NO*	1	0	1
	%	1	0	11.1
MS	*NO*	1	0	0
	%	1	0	0
TOTAL	*NO*	66 (96)	22(22)	5(9)
	%	68.9 %	100%	55.5%

Table 8. Association between the grades of AR and the types of AVS.

Variable		VAS	SAS	SVAS	P value
MILD AR	*YES*	28	10	0	0.041
	NO	68	12	9	
MODERAT AR	*YES*	5	3	0	
	NO	91	19	9	0.246
SEVER AR	*YES*	2	1	0	0.703
	NO	94	21	9	

4. DISCUSSION

In the current study male gender was predominant in all types of aortic stenosis with male/female ratio (1.6:1 _ 3.5: 1).

This statement agreed with the reports of Perry et al. and Rothman and Flyer who reported 64%–78% for aortic stenosis in boys [8,9], and close to Ibn- sena (mousl) teaching hospital report in which 69.2% patients with aortic stenosis were male [10]. Male also is the predominant sex (67% male) in a study has done at Liverpool [11].

We found that most of our patients (44.9%)have presented with an accidental murmur (asymptomatic) eitherduring hospital admission for other reasons (chest infection, gastroenteritis, etc.) or before preparing for surgery. The treating physicians have referred these pts. due to positive auscultatory findings. Features of CHF seen in 33 pts. (26%), 32 pts. of them belong to the valvular type. There was a significant relation between the types of aortic valve stenosis and those pts. Presented with CHF, **p value was 0,004**.Thismight being explained by the severity of VAS as all affected patients were below one year of age.

Others presentations were fatigability, chest pain, shock state due to critical AS and syncope in decreasing frequencies.

A study done in China in which 51 cases with aortic stenosis were included; shows that most of patients (56.9%) were asymptomatic [12]. CHF was noted in 31.2% of cases, exertional dyspnea in (17.6%) of cases, syncope in (9.8%) of cases, and chest pain in (7.8%) of cases.

Other study has done in Toronto; in which 313 cases with AS were conducted, showed that most of the pts.

With aorticstenosis have presented with an accidental murmur (67%) [13].

The most common morphological type of aortic stenosis in our study was valvular aortic stenosis which occurred in 96 children (75.6%) followed by subvalvular stenosis which affect 22 children (17.3%), then supravalvular stenosis affecting 9 children (7.1%).This morphological classification is close to a study done in Liverpool, showed that VAS occurred in 71.2% cases, SAS in13.7%cases and SVAS in 7.7%cases. A study done in china; in which51children diagnosed with aortic stenosis was included, VAS occurred in 39 children (76.5%), SAS in 5 children (9.8%), and SVAS in 7 children (13.7%) [12].

Regarding **VAS,** bicuspid aortic valve was the most common type in the current study, seen in 91 cases (94.8%), tricuspid AV and unicuspid AV founded only in 3 pts. and 2 pts. respectively. These findings were different somewhat from the findings found in a study done in Turkey [14] on 266patients, showed that 127 patients (48%)were bicuspid, tricuspid in 136 patients (51%) and unicuspid in 3 patients (1%).Aortic valve was mostly bicuspid in 371 patients (73%), tricuspid in 107 cases (21%), and unicuspid in 30 cases (6 %) in another study done in Indiana [15].

One single-center study done by Roberts WC and Ko JMincluded 932 patients who underwent aortic valve replacement for AS without mitral stenosis (thus excluding most rheumatic disease) found definite congenital abnormalities in 54% of the aortic valves, with 5% being unicuspid valves and the remainder bicuspid, i.e. in 95% [16].

Regarding **SAS**; sub aortic tissue ridge (membrane) was the most common pathological type in the present study. Thisstatement is in agreement with the report of BrownW et al [15]; in which 132 patients with subvalvular AS were studied, they found 110 cases (83%) have presented with discrete sub aortic membrane and 22 patients (17%) with tunnel type.

Fifty one children with discrete subvalvular aortic stenosis were studied by Newfeld E et al. [17]. The three morphological types of obstruction found were the thin membranous type (43 cases), the fibro muscular collar type (5 cases) and the tunnel type (3 cases).

Regarding patients with **SVAS, t**he cases with hourglass defect were the most common in our study; found in 7 cases (77.8%) while long segment defect documented only in 2 pts. (22.2%).These findings are close to the results of Brown W et al [15]; showed that (72.5 %) of pts. with SVAS had a localized type (hourglass) and (27.5%) of them were belong to the diffuse type and it was also close to the results of van Son et al study in which (83.8 %) of pts. diagnosed with SVAS had localized type and (16.2%) had diffuse type [18].

The most common encountered associated cardiac defect in our study was AR. We found it most commonly in patients with subvalvular AS (68.2%).

Robison R et al study reported that AR is the most common associated defect. They were found it in (59%), (41%), (35%) of pts. diagnosed with subvalvular, supravalvular and valvular aortic stenosis respectively [19].

Regarding the relation between the types of aortic stenosis and the grades of aortic regurgitation, we found that there is a significant association between mild AR and the aortic stenosis type's, ***p value was 0.041***. This may be because that all AR defects were detected early at the time of first presentation.

We found that William's Syndrome was the most common Syndrome documented in the current study and it was presented only inpatients of SVAS. We found a significant association between the types of aortic valve stenosis and the patients diagnosed with William's Beuren Syndrome, p **value was 0.000.**

This statement is close to a study done in France [20], in which they found that Williams Beuren Syndrome was the most common syndrome; sixcases from atotal of 14 cases diagnosed as cases of SVAS were Williams Syndrome. A retrospective Study done in Turkey included files of 24 patients diagnosed with SVAS, 15patients of them were found to have Williams' syndrome [21].

CONCLUSION

1. Valvular aortic stenosis is the most common type of aortic stenosis in this study.

2. Bicuspid aortic valve is the most common congenital anomaly associated with aortic valve stenosis.

3. Most of the patients with aortic stenosis were discovered to have an accidental murmur either by clinical examination before preparing for surgery or during admission to hospital for other reasons.

RECOMMENDATION

1. We recommend a further study to evaluate the prevalence of bicuspid aortic valve in the general population and to compare it to the percentage of the diseased bicuspid aortic valvein our community.

2. We recommend improving neonatal examination at birth in all nursery units for earlierdetection of the serious cardiac defects.

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