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Review

Left ventricular response to pressure afterload in children: Aortic stenosis and coarctation
A systematic review of the current evidence

Haki Jashari [a], Annika Rydberg [b], Pranvera Ibrahimi [a], Gani Bajraktari [a], Michael Y. Henein [a,*]

[a] Department of Public Health and Clinical Medicine, Umeå University, Sweden
[b] Department of Clinical Sciences, Umeå University, Sweden

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ABSTRACT

Congenital aortic stenosis (CAS) and Coarctation of Aorta (CoA) represent two forms of pressure afterload that affect the left ventricle (LV), hence require regular echocardiographic monitoring. Subclinical dysfunction of the LV exists even in asymptomatic patients with preserved left ventricular ejection fraction (EF), implying low sensitivity of EF in predicting optimum time for intervention. In this article we review patterns of LV myocardial deformation before and after correction of CAS and CoA in infants, children and adolescents, showing their important role in monitoring the course of LV dysfunction. A systematic search using PubMed was performed and suitable studies are presented on a narrative form. Normal EF and/or fractional shortening (FS), with subclinical myocardial dysfunction are reported in all studies before intervention. The short-term results, after intervention, were related to the type of procedure, with no improvement or further deterioration related to surgery but immediate improvement after balloon intervention. Long term follow-up showed further improvement but still subnormal function. Thus correction of CAS and CoA before irreversible LV dysfunction is vital, and requires longitudinal studies in order to identify the most accurate parameter for function prognostication. Until then, conventional echocardiographic parameters together with myocardial velocities and deformation parameters should continue to provide follow-up reproducible measures of ventricular function.

1. Background

Congenital aortic stenosis (CAS) and aortic coarctation (CoA) represent two forms of pressure afterload on the left ventricle (LV) due to stenotic valves and/or narrowed aortic root/proximal descending aorta and hence increased backward pressure. Aortic tubal narrowing/obstruction can be isolated or at multiple levels, often in combination with septal defects or conotruncal anomalies, resulting in different forms and severities of pressure and volume LV overload [1]. Congenital AS is commonly (70%) caused by abnormal aortic valve leaflets, although obstruction may affect the subvalvular area (14%) or supravalvular region (8%) or rarely more than one level concomitantly (8%) [2]. The incidence of CAS is 401/million live births, with a clear male predominance 4:1 [3,4]. The commonest leaflet anomaly causing AS is the bicuspid aortic valve disease (BAV), commonly detected in children and adolescents. Unicuspid AS is more frequently seen in neonates presenting with critical AS, and it often requires urgent intervention [5]. The incidence of BAV in the general population is approximately 1–2%, with a male predominance of 2:1 [6,7]. AS caused by BAV syndrome may develop at any age, since the leaflet anomaly makes it prone to calcification and fibrosis, hence, most patients show signs of calcification by the age of 30 years [8].

Discrete coarctation of the aorta consists of short-segment narrowing in the region of the ligamentum arteriosum adjacent to the origin of the left subclavian artery, which may rarely involve the aortic arch or isthmus. Extensive collateral vessels may develop proximal to the obstruction, which may reduce the pressure drop across the CoA and mask the severity of the obstruction. CoA affects 409/million live births with a modest male predominance of 1.5:1 [1,3]. Concomitant aortic lesions with CoA are common, mostly BAV in approximately 50–75% of patients [9].

Aortic obstructive lesions are currently considered not simple anomalies but a genetic disorder of the cardiac and aortic development [10–14]. An autosomal dominant pattern of inheritance has been suggested [15–17]. The 9% prevalence of BAV in first-degree relatives of patients supports the current guidelines of the American College of Cardiology/American Heart Association which recommend echocardiographic family screening [18]. This is further supported by the evidence

* Corresponding author at: Heart Centre and Public Health and Clinical Medicine, Umeå University, Umeå, Sweden.
E-mail address: Michael.henein@medicin.umu.se (M.Y. Henein).

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that considers LV outflow tract and aortic obstructive lesions as intrinsic anomalies of the vascular system, associated with an increase in collagen and decrease of smooth muscle content of the aortic segments proximal to the CoA [12,19] as well as the 53% BAV and 32% of their first degree relatives are likely to develop aortic dilatation [20].

Conventional Doppler echocardiographic techniques are the most commonly used investigations for diagnosing and assessing congenital AS and CoA as well as their effect on the left ventricle. Although ejection fraction has been frequently used to reflect LV systolic function, it is unable to detect associated subclinical function disturbances, particularly at the subendocardial level, commonly seen in pressure afterload pathology, even before obvious myocardial hypertrophy develops [21–24]. Recently MRI has become an attractive non-invasive tool for assessing heart anomalies including congenital AS and CoA but it is known for its limited availability, need for sedation as well as the need for special expertise.

2. Objectives

The objective of this review is to assess LV function changes in infants, children and adolescents with CAS or CoA using various recent echocardiographic modalities including myocardial deformation imaging techniques as well as comparing LV function parameters between pre- and post-intervention among studies.

3. Search engine and evidence criteria

Between July 2014 and August 2014, a systematic search using PubMed for studies reporting LV function in congenital AS and CoA in infants, children and adolescents was performed. Additional studies were identified by a manual search of references. No language or year of publication restriction was applied. Search terms included "aortic stenosis", "valvular stenosis", aortic coarctation, left ventricular, "left ventricular dysfunction", "left ventricular function", infants, children, adolescents, pediatric, "two dimensional speckle tracking echocardiography", "tissue Doppler imaging", "left ventricular twist", "left ventricular rotation", "left ventricular torsion", "left ventricular strain", "strain of the left ventricle", "torsion of the left ventricle", "twist of the left ventricle" and "rotation of the left ventricle".

3.1. Inclusion and exclusion criteria

We included studies published between 2006 and 2013. Two researchers (HJ, PI) independently reviewed titles and abstracts of the search results, including only studies that compared post-intervention to pre-intervention LV function in infants, children and adolescents with congenital AS and/or CoA. Editorials, review articles and case reports were excluded. Full text articles were then retrieved and reviewed. Reference lists of the retrieved articles were searched manually for potentially relevant articles.

3.2. Data extraction

One reviewer extracted the data and summarized the findings of the studies. Predetermined data of interest were the first author, year of publication, title, journal, study population, age of population at time of intervention, type of intervention, duration of follow-up, presence or absence of a control group, method of LV function evaluation and LV function parameters.

3.3. Data synthesis

Statistical meta-analysis of the results was unsuitable due to the different methodologies used in individual studies. However, we decided to present the results in a narrative form, dividing parameters of LV function into, pre-intervention, early post-intervention (up to 6 months) and late post-intervention (beyond 6 months).

4. Qualified studies

The PubMed search identified 4945 articles. After a review of titles and abstracts we identified 136 papers reporting LV function in congenital AS and CoA in the pediatric population. Having critically reviewed the full text, only 7 papers proved suitable for inclusion in this review (Fig. 1 and Table 1). Four, one and two studies reported on congenital AS [25–28], CoA [29] and combined diseases [30,31], respectively. Three studies reported on LV function pre-, early post- and late post-intervention intervals [26,28,31]. Two studies reported LV function only on pre- and early post-intervention [25,30], while the remaining two studies reported only on pre- and late post-intervention LV function [27,29]. Four studies evaluated LV function with TDI [25,27–29], three of them used GE EchoPAC [25,28,29] as offline analysis software, and in the forth study the TDI analysis tool was not reported [27]. Offline speckle-tracking analysis was performed using GE EchoPAC software in three studies [26,30,31]. The total number of studied patients was 199, 123 with CAS and 76 with CoA. Four studies included a total number of 188 healthy controls. All studies reported normal LV systolic function in the form of EF and/or fractional shortening (FS) pre-intervention.

4.1. Aortic stenosis

Patients with CAS requiring intervention had already clear evidence for left ventricular long axis dysfunction before intervention [26,30,31] while EF and/or FS were within normal ranges. Marcus et al. [26] reported simultaneously reduced circumferential and radial strain/strain rate parameters together with suppressed longitudinal function. However, Laser et al. [30] reported an accentuated circumferential function as well as enhanced torsion preoperatively as potential compensatory mechanisms to the disturbed longitudinal function.

The short-term effect of CAS repair seems to be dependent on the type of intervention used. Balloon aortic valvuloplasty (BAVP) resulted in significant improvement of LV function within hours [30] or days [25] compared to pre-intervention but still was subnormal six months post-BAVP [26]. On the other hand surgically treated patients had no improvement of global strain [28] but rather further deterioration of longitudinal strain [31] a few weeks after surgery.

Long-term improvement of global [28] and longitudinal strain [31], despite remaining subnormal, has been reported. Likewise, Marcus et al. [26] noticed significant improvement of longitudinal, radial and circumferential strain/strain rate, which still remained subnormal, despite the normalization of radial function.

LV diastolic function has also been found to be abnormal in CAS patients, and is influenced by intervention. While balloon aortic valvuloplasty leads to short [25] and long term [27] improvement of LV diastolic function, Mi et al. [28] reported non-significant improvement 12 months after surgical aortic valve replacement (AVR).

4.2. Coarctation of Aorta

LV systolic dysfunction, in terms of myocardial deformation parameters, has been reported in patients with CoA prior to intervention [29–31]. LV longitudinal strain has been found decreased but to a lesser extent compared to CAS [30,31], with the circumferential strain/strain rate and torsion being higher than in CAS [30].

As is the case with CAS, short-term LV response to intervention was dependent on the procedure used. LV maximum torsion reduced soon (4–6 h) after balloon aortoplasty, despite the need for additional stenting in almost two thirds of patients [30]. In contrast, Van der Ende et al. [31] reported further longitudinal strain reduction, one week post-intervention irrespective of procedure.