PITFALS IN DIAGNOSIS, MANAGEMENT AND TREATMENT OF HYPERTENSION DUE TO LATE PRESENTING CoA

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ABSTRACT

Introduction:

Coarctation of the aorta (CoA) is common Congenital Heart Disease presenting by many symptoms/signs, in any age group. From Hypoplastic Left Heart Syndrome, at birth, as Heart Failure in infancy, to undetected Hypertension (HTN) even in late adulthood. HTN, a major co-morbidity presenting in any age, even after surgical treatment. It’s difficult to control by pharmacotherapy. Intracardiac cerebral complications of uncontrolled HTN are basic reasons for patients to have long-life follow-up and increasing morbidity/mortality. The so-called golden standard of absent/diminished femoral artery pulsation and cardiac murmurs can be absent or not picked-up in 40%.

Aim: presenting our experience on diagnosis, management and treatment of HTN in late CoA presenters.

Population:1031 children - adolescents were referred to our clinic for investigation of atypical cardiac complains, during the last six years by their pediatricians. Age ranged: 4.5 - 17.1 years old (mean age:6.9 years).

Method: Diagnosis based: medical history, clinical examination, Echo-2D scan, 24h BP Holter. CT, MRI, catheterization confirming diagnosis.

Results: 116 diagnosed with late CoA. 25/116(21.6%) was suspected HTN and 16/116(13.8%) were thought to be CoA by their pediatricians.

Physical findings of CoA:

Presented with :

- ❖ 68/116 (58.6%), had a murmur and reduced femoral pulses.
- ❖ 23/ 116 (19.8 %) had only HTN
- ❖ 25 /116(21.55%) had also additional HTN.

Treatment:

- 94/116 were treated surgically. Restenosis occurred in 35.05%.
- 12/116 were treated by interventional. Restenosis occurred in 40.95%
- * 18/116(15.52%) patients had none /trivial restenosis but presented HTN during the F/U period.
- ** 9/48 initial patients were normotensive after surgical/intervention treatment.

Medicine guidelines treatment: 48/116 had HTN and were F/U for a mean period of 54 months.

Treatment compliance: 80.40%.

- ❑ 28/57 (49.12%) successful control HTN
- ❑ 10/57 (17.54%) poorly controlled with 2 medications (ACE-I, b-Blocker)
- ❑ 18/57 (33.33%) poorly controlled even with 3 medications(ACE-I, b-Blockers, CaChB)

Conclusions: CoA was mist by pediatricians in 64.65% of its late presenters. A 15.52%, secondary HTN even after successful surgical treatment was noticed. When HTN is present at least 1/3 will need a combination of two or more medicines to control their BP and 1/3 of the HTN patients will be poorly controlled. Further, larger in cohort size and longer F/U periods, prospective studies are needed, to verify our findings.

INTRODUCTION

CoA is a narrowing of the lumen of the isthmus, adjacent to the Ductus Arteriosus (DA) on the posterior wall of the thoracic aorta. It is an angiopathy of the Aorta1. It is the fifth most common type of CHD, found in approximately 1/2500 live births2 with a favorable prevalent in males. This frequency underestimates delayed diagnosis, even in the adult population, due to many late presenting cases3. It can also create intracranial aneurysms4. Clinical, presents in any age. In Fetal as a part of Hypoplastic Left Heart Syndrome(HLHT), to early neonatal life, at the end of the first week of life when the DA closes. It presents, as a Low Cardiac Output Syndrome(LCOS) with Congestive Heart Failure (CHF), acidosis, acute renal failure and early death, that mimics late neonatal sepsis1,5. In cases of favorable anatomy, these can present in any age from childhood to adulthood either during an investigation of a murmur or due to incidental diagnosis of HTN. Frequently we find the combination of a complex CHD that involves a perimembranus ventricular septal defect [(pm) VSD] with a bicuspid aortic valve (BAov) that can be normal or have any amount of stenosis(AovS) or regurgitation1,5. CoA is present in 18% of patients with Turner’s6 and in 10% of patients with William’s syndrome7,8.Is also present in Shone’s complex1,5,7,8. Less frequent but more tragic presentation, can be a rapture of intracranial aneurisms that can be found in 5-10% of these patients8,9. The diagnosis is based on the combination of medical history, physical examination and imaging techniques5. Investigation of patients with accidental findings of “new” systolic or continues murmurs (collateral vessels around the CoA), mostly in the intrascapular area, as well as the detection of HTN with a reverse phenomenon in which the BP in the upper, is higher than in the lower extremities, by at least 20mmHg. These findings with additional diminished or delayed femoral arterial pulses has been thought in the past to be the gold standard findings that points directly to CoA5. Newly studies show that these are present in 60% of patients suffering from CoA10. Additional evidence can be added by a Chest X-Ray. Electrocardiography (ECG) can demonstrate in late onset, evidence of left ventricular hypertrophy (LVH). Less invasive, cheap and easy to provide, today is Echocardiography (Echo-2D)10.

Cardiac catheterization, today has limited diagnostic value, but I important in interventional management8,10. Magnetic Resonance Imaging (c-MRI) Computer Tomography (cv-CT) are frequently used. Gadolinium-enhanced c-MRI angiography/ 3D reconstruction images, provides excellent data. Analysis through phase contrast flow estimates the gradient across the CoA11. The use of cv- CT must be considered in selected patients, not c- MRI compatible12-13.

Today, we offer both **interventional** and/or **surgical** options of treatment14,15. A native CoA or re-CoA, with a gradient greater of 20 mm Hg, is an indication for treatment. Longstanding native CoA with significant collateral flow, have a lower measured gradient despite severe CoA16.Management is based on the age of presentation, complexity of anatomy, and whether the CoA is a native vs recurrent obstruction. In very early presenting native CoA, surgical repair is preferred 17. BA is a palliative in critical neonates considered too sick for surgery8. Surgery is appropriate in patients with complex CoA 16. In older patients, with a simple, small, juxtaductal, native CoA, stent placement is a reasonable approach14. For recurrent CoA, we consider initial BA17,18. Finally, treating resistant early or late HTN, even after successful surgical treatment is a burden. This difficult to control by pharmacotherapy, and the late complications of uncontrolled HTN, are the principle reasons for these patients to have a long-life follow-up. This population, while aging, will face important and increasing morbidity and mortality8.

AIM

Presenting our experience on diagnosis, management and treatment of HTN in late CoA presenters.

POPULATION-METHODS

1031 patients aged: 4.5 – 17.1 years old (mean age 6.9 years) were examined for different reasons in 6 centers in Baku, Azerbaijan, during the period 08/2012-08/2018.

116(11.25%) were detected to suffer from late CoA.

Method: Diagnosis based: medical history, clinical examination, Echo-2D scan, 24h BP Holter. CT, MRI, catheterization confirming diagnosis. Mean follow-up period 55 months.

RESULTS

116 diagnosed with late CoA. 25/116(21.6%) was suspected HTN and 16/116(13.8%) were thought to be CoA by their pediatricians.

Physical findings of CoA:

Presented with :

- ❖ 68/116 (58.6%), had a murmur and reduced femoral pulses.
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Treatment:

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Treatment compliance: 80.40%.

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DISCUSSION

CoA is a common CHD8. Despite early presentation in neonatal life, many cases can be mist if BP is not obtained in each well baby visit after the age o3 years old as the AAP guidelines suggest20. In longer-term anti-HTN therapy is required as chronic HTN, can follow even successful treatment with no residual gradient. Anti-HTN medicines must be used aggressively to maintain normal BP. In 2008 the ACC/AHA Guidelines for the Management of Adults with CHD recommend use of an ACE-i, as first-line therapy. Additional treatment with b-blockers, Calcium channel blockers and Angiotensin II receptor blockers can be used. When monotherapy is not successful -most of the cases of late CoA treatment- dual or even three classes of anti-HTN medicines must be used to normalize BP21.As HTN can be seen early or late after even a successful treatment a lifelong prevention strategy is needed. Studies have shown that 30% of ex-CoA treated patients will suffer from HTN by adolescence raising to about 60% in adulthood. This suggests that HTN may be an inevitable consequence of CoA, even when an effective repair has been achieved early in life. For this reason, late detected patients will suffer from chronic HTN. Despite the progress in pharmacotherapy the best medicine treatment is debatable and will include more than one class of anti-HTN drugs. Primary prevention, will be as soon as possible to detect and early operate before the first year of life21,22. Secondary prevention aims to eradicate any residual stenosis and treat existing HTN if present with an ACE-i or if not controlled on monotherapy to add other anti-HTN classes22-23. Finally, tertiary prevention would be treating aggressively, lifelong HTN 22-23. Further research into the mechanisms leading to HTN may identify therapies to target the vasculopathy changes seen in CoA and improve outcome21-25.

CONCLUSION

We concluded that CoA was mist by pediatricians in 64.65% of its late presenters. A 15.52%, secondary HTN even after successful surgical treatment was noticed. When HTN is present at least 1/3 will need a combination of two or more medicines to control their BP and 1/3 of the HTN patients will be poorly controlled. Further, larger in cohort size and longer F/U periods, prospective studies are needed, to verify our findings.

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