

CARDIOMAG NEWSLETTER

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Dear Doctor,

Greetings from Micro Labs Limited. We are happy to bring you “Cardio Mag” which contains latest and interesting news in the field of Cardiology

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Study of Thoracic aortic dilatation, aortic dimensions and growth, and the incidence of aortic dissection in women with Turner syndrome: A Systematic review

Turner syndrome (TS) or congenital ovarian hypoplasia syndrome is one of the common sex chromosomal abnormality found in females.¹ TS affects women with both absent or incomplete X-chromosome. Approx. 1:2500 is the incidence of TS among live-born females. However, actually prevalence rate is unknown as many patients may remain undiagnosed. Occurrence rate of this syndrome similar in different ethnicities and different countries. Common features of TS includes short stature, gonadal dysgenesis, and cardiovascular disease.² Common congenital cardiac defects seen in Turner syndrome include bicuspid aortic valve (BAV), aortic coarctation (COA), abnormal pulmonary venous return, aortic dilatation and dissection. However, at the end of the 20th century, increased in the case reports published on aortic dissection in women with TS raised the awareness and research on aortic disease.¹

Meccanici F et al aimed to review and analyse the prevalence of thoracic aortic dilatation, aortic dimensions and growth, and the incidence of aortic dissection in patients with Turner syndrome

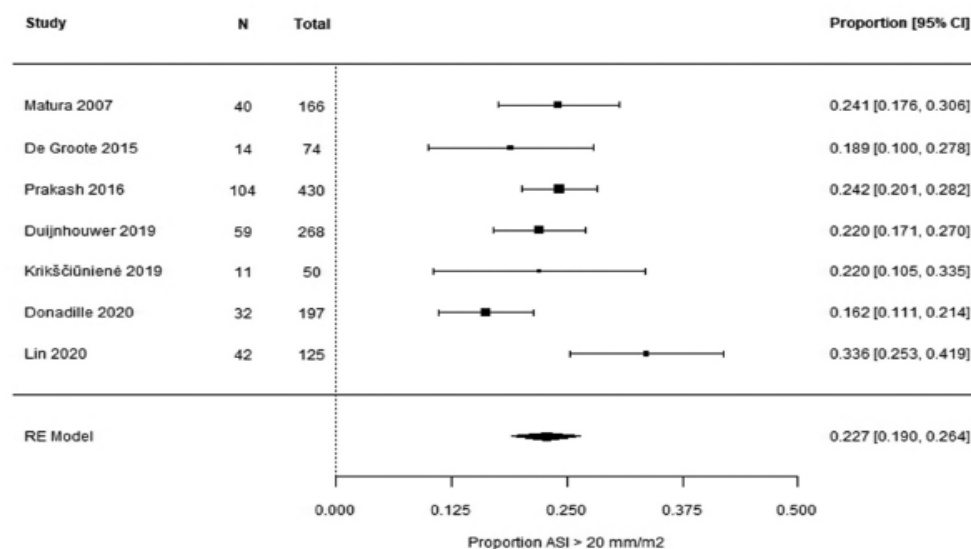


Figure 1: Forest plot of prevalence of aortic dilatation defined as an ASI > 20 mm/m2 in the ascending aorta.

In this analysis, systematic literature searches were done and observational studies with diagnosed Turner syndrome were included, while studies with children aged <15 years old or specific Turner syndrome were excluded. 21 studies were screened and included. 23% was the pooled prevalence of ascending aortic dilatation (fig 1) while the incidence of aortic dissection was 164 per 100.000 patient-years. However, 3 studies showed aortic growth over time to be limited. It was also observed that the risk factors for aortic dilation were increased age, bicuspid aortic valve, aortic coarctation, and hypertension.

Conclusion: This study concluded that, ascending aortic dilatation is common in adult women with Turner syndrome and the hazard of aortic dissection increases too when compared to general population, and aortic growth is limited. It is not all possible for Conventional risk markers to identify and explain all aortic dissection cases, that is why new imaging parameters and blood biomarkers are required to improve prediction, making easy patients follow-up and surgical decision-making

Refereneces

1. Meccanici F,etal. Prevalence and development of aortic dilation and dissection in women with Turner syndrome: a systematic review and meta-analysis. Expert Review of Cardiovascular Therapy. 2023 Feb 1(just-accepted)
2. Shankar Kikkeri N, Nagalli S. Turner Syndrome. [Updated 2022 Aug 8]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2022 Jan-

Cardiovascular assessments and management of findings in patients with Eating Disorders

Eating disorders (EDs) are defined as the serious conditions with marked imbalances in eating behaviour and are also associated with distorted body image and extremes of weight. Eating disorders is commonly observed in children, adolescents and young adults that considerable threat to their health and wellbeing leading to severe psychiatric and medical complications and decreased quality of life.¹ Eating disorders is classified into four well defined disorders including binge eating disorder (BED), avoidant/ restrictive. Eating disorders must be immediately diagnosed and treated to avoid high morbidity and mortality associated with this condition²

According to studies, 1 in 8 young adults may have at least one eating disorder by twenty years of age. Prevalence of anorexia nervosa and bulimia nervosa are approximately 0.3% and 1% among adolescent females respectively.²

Cases of eating disorder was found to be increased in all age groups following the Covid-19 pandemic with patient's presentations being more medically compromised compared to previous years. This study aimed to study and identify the physiological context in which cardiovascular complications develop, systematically outline these complications and to suggest a pragmatic approach to their clinical evaluation in people with eating disorders. To conduct this review, relevant literature, guidelines and academic texts were studied and analysed. Conclusions of each studied were extracted and were verified by a Child and Adolescent Psychiatrist and Adolescent Paediatrician before adding to the study. This study also highlighted that the cardiovascular complications in Eating disorders are mostly associated to malnutrition, and patients having Anorexia Nervosa (fig 2) have higher chances of structural and functional cardiac abnormalities, such as aberrations of heart rate and rhythm, haemodynamic changes and peripheral vascular abnormalities.

Structural and functional cardiac abnormalities	Repolarisation and conduction abnormalities	Haemodynamic abnormalities	Peripheral vascular abnormalities
Decreased left ventricular mass (LVM) Decreased left ventricular systolic function Mitral valve prolapse Pericardial effusions Myocardial fibrosis → Increased risk of sudden cardiac death ←	Sinus node dysfunction: Sinus* bradycardia, Sinus pauses and Junctional escape rhythms AV conduction delay QTc prolongation and Increased QT dispersion (secondary to electrolyte deficiency and medications)	Hypotension* Orthostatic hypotension and tachycardia* Syncope* Autonomic dysfunction	Acrocyanosis Dysregulation of peripheral vasoconstriction and vasodilation

Figure 2: Summary of cardiovascular complications in anorexia nervosa

In most patients with Bulimia Nervosa, cardiovascular abnormalities are observed secondary to electrolyte imbalances. Avoidant or restrictive Food Intake Disorder and Binge Eating Disorder are commonly associated with distinct cardiovascular complications. With nutritional restoration, and normalisation of eating behaviours, some of the cardiovascular complications get reversible, such as cessation of purging. Complete and thorough physical examination along with detailed clinical enquiry is required to ensure the medical safety of young adults with eating disorder.

Conclusion: Analysing cardiovascular Complications in Eating disorder and collaborating with acute medical teams allows community clinicians to detect patient at highest risk of and minimise the complications

References

1. Friars D, Walsh O, McNicholas F. Assessment and management of cardiovascular complications in eating disorders. *Journal of Eating Disorders*. 2023 Dec;11(1):1-2
2. Balasundaram P, Santhanam P. Eating Disorders. [Updated 2022 Sep 3]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2022 Jan-. Available from: <https://www.ncbi.nlm.nih.gov/books/NBK567717/>

Evaluation of the diagnostic performance of a method for acoustic analysis of turbulent murmur caused by coronary artery stenosis in patients with coronary artery disease

Cardiovascular disease has not only increased economic burden but also has threatens human health and life. According to studies, approximately 330 million people have cardiovascular diseases including 11.39 million people with coronary artery disease. coronary artery atherosclerosis is one of the common cause of coronary artery disease leading to myocardial ischemia, hypoxia, or necrosis. However, some studies have come up with an acoustic analysis of turbulent murmur caused by coronary artery stenosis (a new and promising method for the diagnosis coronary artery disease). Dock et al. by using phonocardiography had detected a diastolic murmur associated with coronary stenosis. After sometimes, other studies too reported heart murmurs similar to Dock's murmur. In last few years, diastolic murmur signal associated with coronary artery stenosis has been detected with modern acoustic detection equipment and analysis methods

Zhao P G aimed to evaluate the diagnostic performance of a method for acoustic analysis of turbulent murmur caused by coronary artery stenosis in coronary artery disease (CAD). This prospective study enrolled 452 Patients admitted to the cardiovascular department for elective coronary angiography. For recording heart sounds a digital electronic stethoscope was used before angiography. Quantitative coronary angiography (QCA) was used as a "gold standard" for the diagnosis of Coronary artery disease and to evaluate the diagnostic performance of the acoustic analysis method for Coronary artery disease

A total of 452 patients completed Quantitative coronary angiography and heart sound data. Acoustic analysis revealed 310 disease cases and 142 normal cases. Increasing the values of coronary artery diameter stenosis from 30% to 50%, 70%, and 90% raised the sensitivity and negative predictive value of the acoustic analysis method. the sensitivity was 75.6%, 81.9%, 83.3%, and 85.7%, respectively while the negative predictive value was 33.1%, 57.0%, 69.7%, and 88.0%, respectively. On the other hand, specificity and positive predictive value decreased i.e. 75.8%, 70.4%, 51.0%, and 37.5%, respectively and AUC values were 0.757, 0.762, 0.672, and 0.616, respectively.

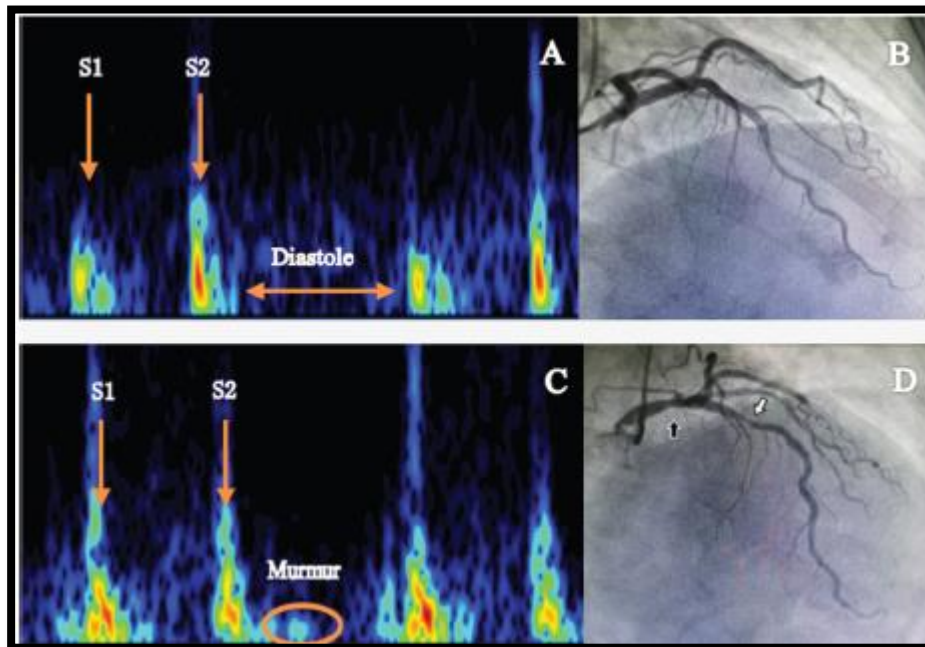


Figure 3:Acoustic Analysis Results and Corresponding Angiograms

It was also found that the sensitivity of the acoustic analysis method for one-vessel disease was 86.6%. At the same time, sensitivity for identifying left anterior descending coronary artery lesions was at 90.7%. The sensitivity for identifying isolated coronary artery branch lesions was 66.7% and for identifying three-vessel disease in multi-vessel coronary artery lesions was at 82.9%.

Conclusion: For the diagnosis of Coronary artery disease, acoustic analysis of turbulent murmur has favourable performance in patients with coronary artery stenosis

References: Zhao PG,etal. Diagnosis of Coronary Artery Disease by Acoustic Analysis of Turbulent Murmur Caused by Coronary Artery Stenosis: A Single Center Study from China. Cardiovascular Innovations and Applications. 2023 Jan 31.

Cardiac transplantation related hemochromatosis.

A case report

Hemochromatosis is defined as the excessiveness storage of the iron. Autosomal recessive disorder associated with a mutation of the *HFE* gene located on chromosome 6 is characterized as Primary or hereditary hemochromatosis. Incidence rate of Primary or hereditary hemochromatosis is of 0.1% to 0.5%, while Secondary hemochromatosis occurs as a consequence of iron overload caused by other complications (anaemia, repeated blood transfusions, long-term haemodialysis, or chronic liver disease). Later, predisposes to iron overload cardiomyopathy, eventually leading to end-stage heart failure, requiring an advanced management, such as heart transplantation.

A 10 -year-old, female patient was presented with the diagnosis of heart failure, uncontrolled hyperglycaemic metabolic state, and acute renal failure. Patient had complaint of acute abdominal pain, progressive orthopnea for the past 48 hours, and decreased diuresis. Past history included diagnosis of type I diabetes mellitus, with ketoacidosis 3 months prior. Physical examination showed hyperpigmented macules in the skin. Patients had tachypnea, with baselinem crepitant rales and hyperdynamic precordium. ECG showed left bundle-branch blockage, cardiomegaly at the expense of the left ventricle (LV). Bilateral pleural effusion, and increased pulmonary flow in the chest X-ray were also noticed. Echocardiogram also revealed a dilated vena cava,(fig 4) moderate mitral regurgitation with left ventricular ejection fraction (LVEF) of 15%.Later, magnetic resonance imaging (MRI) revealed global hypokinesis with greater involvement of the apical and mid inferolateral and anterior wall of the LV, global hypokinesis of the right ventricle, LVEF of 17%, right ventricular ejection fraction of 21%, biventricular dilated cardiomyopathy, patchy enhancement, with epicardial and endocardial involvement at the inferoseptal and lateral level at the mid LV.

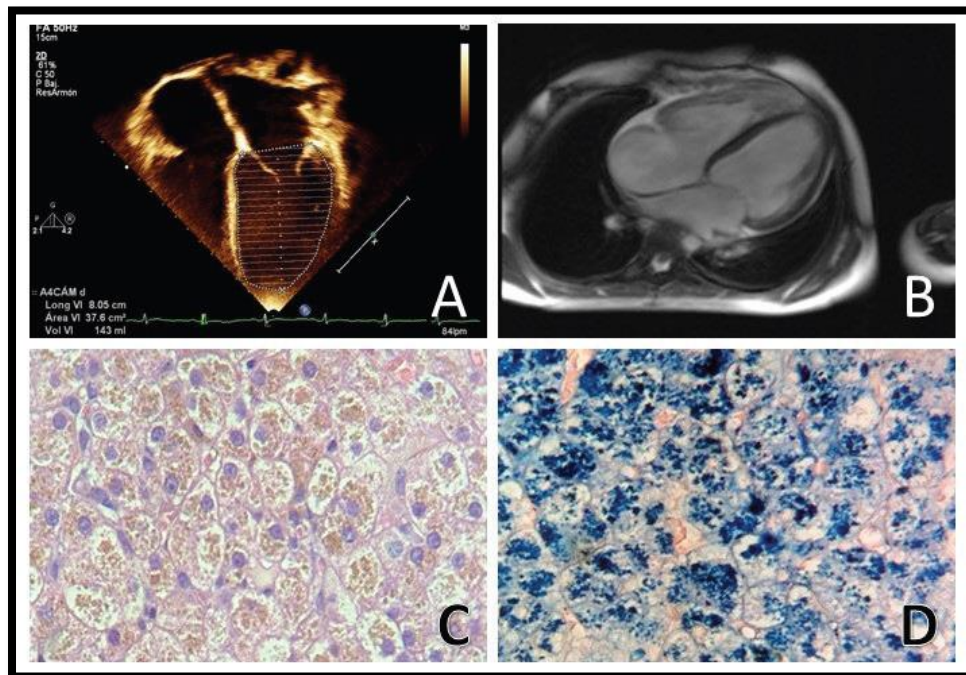


Figure 4:. (A) Echocardiogram showing cavity dilation. (B) Magnetic resonance imaging showing biventricular dilated cardiomyopathy. (C) Hepatocytes with abundant golden-brown pigment, hematoxylin and eosin staining. (D) Positive Perls' Prussian blue stain

Viral panel showed negative and an infectious myocarditis was diagnosed. To confirm, hemochromatosis, a test protocol was initiated. Needle biopsy of the liver highlighted fibrosis, increased inflammatory infiltrate without activity, and enlarged hepatocytes with abundant golden-brown pigment in the lobule, positive for the Perls' Prussian blue stain. Iron chelation therapy and phlebotomy was initiated. patient was discharged and was re-admitted for several times due to refractory heart failure, for which was chosen for heart transplantation. After 7 months' orthotropic heart transplantation using the bicaval approach was conducted. Post-transplantation, echocardiogram showed a LVEF of 65%. Fibroelastosis and cardiomyocyte hypertrophy, with deposits of coarse granules of hemosiderin were the findings of microscopic examination.

A week later, post-transplantation endomyocardial biopsy showed acute mild rejection according to the grading criteria for acute cellular rejection of the International Society for Heart and Lung Transplantation. Patient was discharged and follow up data revealed that patient continued with iron chelation therapy based on deferoxamine and test values reduced sharply and progressively in repeated iron kinetics assessments. Four years later, iron kinetics values were normal and patient was clinically improved.

Conclusion: This case report highlights the multidisciplinary approach needed for the diagnostic and therapeutic process of a patient with hemochromatosis

Reference: Contreras-Ramos A, Bolio A. Cardiac transplantation due to hemochromatosis. A pediatric case report. Arch Argent Pediatr. 2023:e202202775.



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