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RESEARCH ARTICLE

EXPLORING THE UNCOMMON: A PICTORIAL JOURNEY THROUGH RARE CONGENITAL HEART DISEASES VIA CARDIAC CT IMAGING

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Abstract

This pictorial essay embarks on an illuminating expedition through the realm of rare congenital heart diseases, guided by the lens of cardiac CT imaging. Through a meticulously curated collection of images, it unveils the intricate nuances and peculiarities of these uncommon cardiac conditions, offering unparalleled insights into their anatomical variations, diagnostic challenges, and clinical implications. With each vivid portrayal, the essay elucidates the diverse spectrum of anomalies, from the esoteric to the extraordinary, showcasing the power of advanced imaging techniques in unravelling the mysteries of the human heart. As a visual odyssey through the corridors of medical rarity, this pictorial journey not only enriches our understanding of congenital heart diseases but also underscores the indispensable role of cardiac CT imaging in their comprehensive evaluation and management.

Introduction:-

Congenital heart diseases (CHDs) present a multifaceted challenge in clinical practice, encompassing a spectrum of structural abnormalities affecting the cardiovascular system. As the most prevalent type of birth defect, CHDs account for a significant portion of paediatric morbidity and mortality worldwide [1]. While advancements in medical imaging have revolutionized the diagnosis and management of CHDs, the complexity of these conditions necessitates ongoing exploration of novel diagnostic modalities to enhance patient care outcomes.

In recent years, cardiac computed tomography (CT) has emerged as a cornerstone in the diagnostic armamentarium for CHDs [2]. With its ability to provide high-resolution, three-dimensional images of cardiac structures, cardiac CT offers unparalleled insights into the anatomical intricacies of CHDs, facilitating precise characterization and therapeutic planning [3]. Moreover, cardiac CT enables comprehensive assessment of cardiac function, vascular anatomy, and hemodynamicparameters, thereby contributing to a holistic understanding of CHD pathophysiology.

This pictorial essay aims to elucidate the radiological aspects of complex congenital heart diseases (CHDs) through 20 cases utilizing cardiac computed tomography (CT) imaging. Cardiac CT has emerged as a valuable non-invasive imaging modality for the assessment of CHDs, offering high spatial resolution and detailed anatomical visualization. Each case presented in this essay showcases the utility of cardiac CT in delineating intricate cardiac structures, identifying abnormalities, and aiding in pre-procedural planning. Through comprehensive analysis and discussion, this essay seeks to enhance the understanding of cardiac CT's role in the evaluation and management of complex CHDs among healthcare professionals.

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The aim of this pictorial essay is to:
1. To demonstrate the capabilities of cardiac CT imaging in the evaluation of complex congenital heart diseases through a series of 20 cases.
2. To highlight the superior spatial resolution and detailed anatomical visualization provided by cardiac CT for the assessment of cardiac morphology and pathology.
3. To illustrate the use of cardiac CT in identifying specific anatomical abnormalities, such as ventricular septal defects, atrial septal defects, and complex cardiac malformations.
4. To provide educational material for healthcare professionals involved in the interpretation and utilization of cardiac CT imaging in the evaluation of congenital heart diseases.
5. To stimulate further research into advanced cardiac CT techniques and protocols aimed at optimizing imaging quality and diagnostic accuracy in patients with congenital heart diseases.
6.To serve as a valuable resource for clinicians involved in the care of patients with complex CHDs, aiding in clinical decision-making and improving patient outcomes through the optimal utilization of cardiac CT imaging.

Research Methods And Design:-

1. Setting:

The study is conducted at a tertiary care academic medical centre equipped with state-of-the-art imaging facilities and specialized expertise in paediatric and adult congenital cardiology. This setting offers access to a diverse patient population spanning different age groups and CHD complexities, ensuring the inclusion of a broad spectrum of cases for analysis. The advanced imaging capabilities of the institution facilitate high-quality cardiac CT scans, essential for accurate radiological assessment and interpretation.

2. Case Identification and Collection:

- Cases were identified from the institutional database of cardiac CT scans performed for the evaluation of congenital heart diseases.
- A total of 20 cases were selected to represent a diverse spectrum of complex congenital heart diseases, ensuring adequate coverage of different anatomical and pathological variations.

3. Imaging Protocol:

At our facility, we conduct 128-MDCT scans using a GE Siemens machine, adhering to meticulous protocols, and employing advanced image analysis techniques. A standardized procedure involves the automated intravenous injection of an 80-mL contrast bolus at a rate of 5 mL/s, followed by a 60-mL saline chaser. Imaging acquisition parameters include a collimation of 32 × 0.6 mm, section acquisition of 64 × 0.6 mm, gantry rotation time of 330 milliseconds, pitch ranging from 0.20 to 0.43, tube voltage set at 120 kV, and a maximum tube current of 400 mAs/rotation. Subsequent image reconstruction is critical for the comprehensive evaluation of congenital heart disease (CHD) patients. While axial images form the basis for diagnosis, multi-planar reconstruction allows extensive analysis, particularly for atrioventricular septal defects. Maximal intensity projection is vital for delineating great vessels, with adaptable slab thickness aiding in scrutinizing aortic arch and pulmonary artery anomalies. Additionally, volume rendering is instrumental in pre-surgical assessment, offering enhanced 3D anatomical depictions surpassing catheter angiography capabilities. Meticulous optimization of CT window and level settings elucidates intra-cardiac shunts with clarity, facilitating comprehensive diagnostic evaluation and pre-procedural planning.

Results/ Case Series:-

- Each case is presented sequentially in the pictorial essay, with detailed descriptions of the congenital heart defect and imaging findings.
- Images are annotated to highlight key anatomical structures, abnormalities, and diagnostic features.

Case 1:

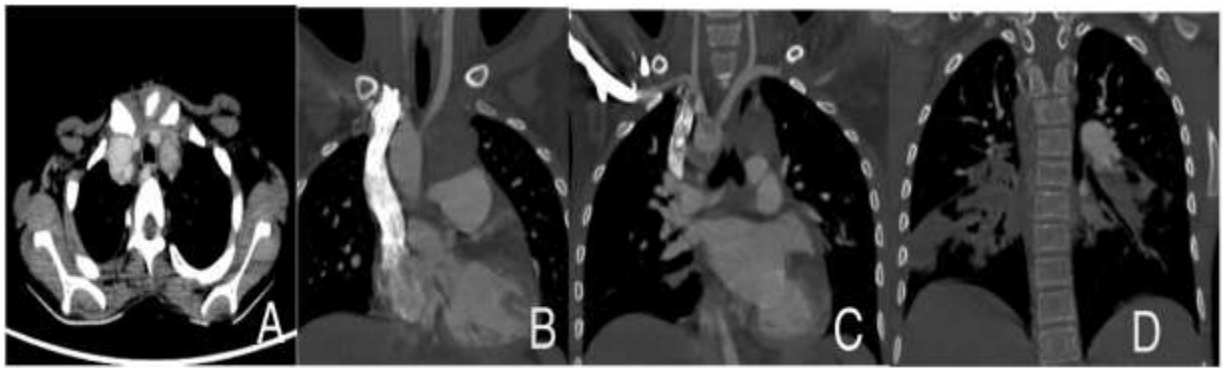

Figure 1:- 11-year-old with tetralogy of fallot. A) Axial CT image demonstrating the retrotracheal course of the left subclavian artery in an 11-year-old female with Tetralogy of Fallot. B) And C) Illustrating separate origins of bilateral common carotid arteries and bilateral subclavian arteries, respectively, leading to vertebral arteries. D) Coronal CT image revealing thrombus formation within the branches of the left pulmonary artery.

Case 2:

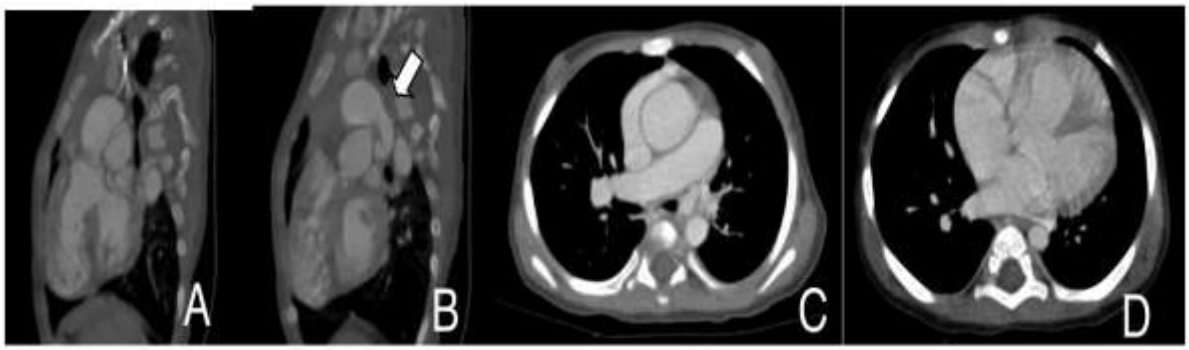

Figure 2:- 1 year old female with history of repeated upper respiratory tract infections was brought with breathlessness and inability to thrive. A - Oblique CT image displaying a Ventricular Septal Defect (VSD) with overriding of the arch of the aorta. B- Depicts a tubular Patent Ductus Arteriosus (PDA). C- Shows a dilated right pulmonary artery and a hypoplastic left pulmonary artery. D- Axial CT image depicting the Secundum type of Atrial Septal Defect (ASD).

Case 3:

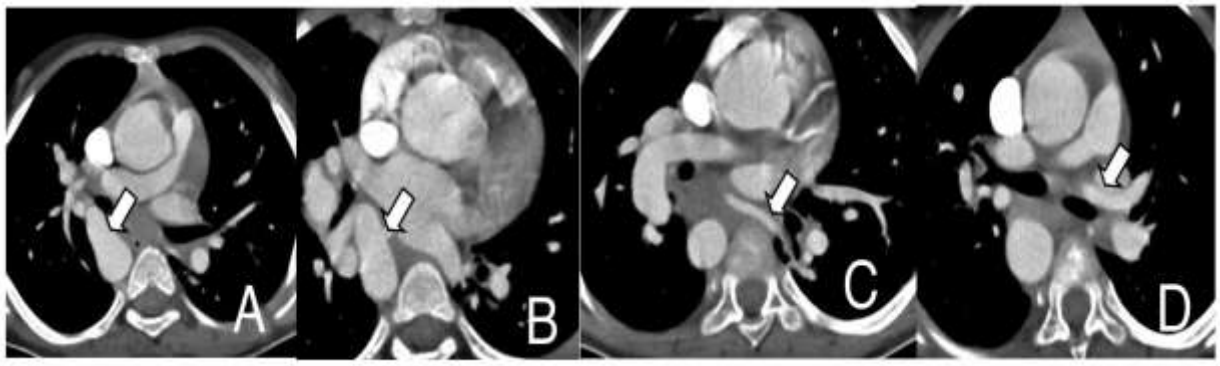

Figure 3:- 5-year-old female with easy fatiguability, breathlessness and inability to gain weight.A and B - Axial CT image showing communicating type of MAPCAs arising from descending aorta at level D6 at 10 O'clock position and at level D7 at 11 O'clock position. C and D show non communicating MAPCAs i.e., directly supplying lung parenchyma and not through pulmonary branches.

Case 4:

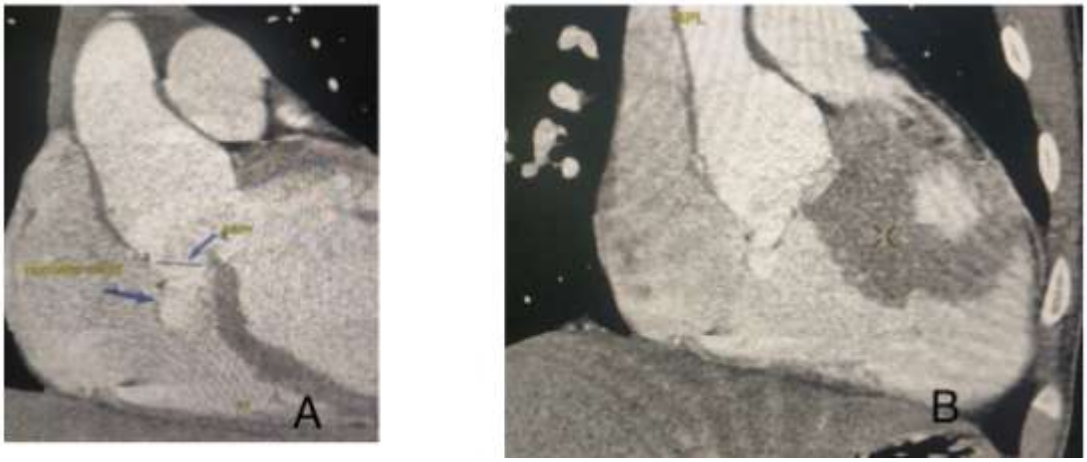

Figure 4:- 14-month-old male child with chest complaints. Oblique post-contrast cardiac CT revealing a fistulous tract originating from the aortic sinus, opening into the right ventricle just below the tricuspid valve, indicative of a ruptured sinus of Valsalva.

Case 5:

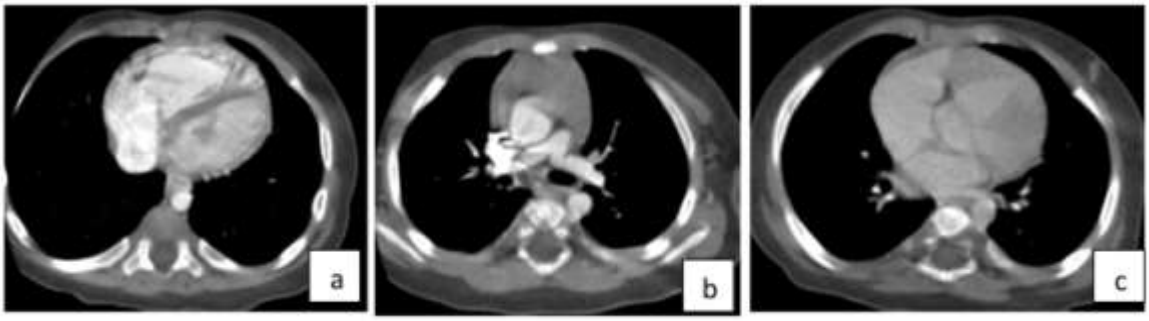

Figure 5:- 6-month-old male with difficult breathing. Axial post-contrast cardiac CT images illustrating: a) the morphological right ventricle positioned on the right side of the morphological left ventricle (D-loop). b) Absence of the main pulmonary artery with the presence of right and left pulmonary arteries suggestive of type 1 pulmonary atresia (by Somerville classification of pulmonary arterial anatomy in pulmonary atresia with ventricular septal defect). c) Normal drainage of pulmonary veins into the morphological left atrium.

Case 6:

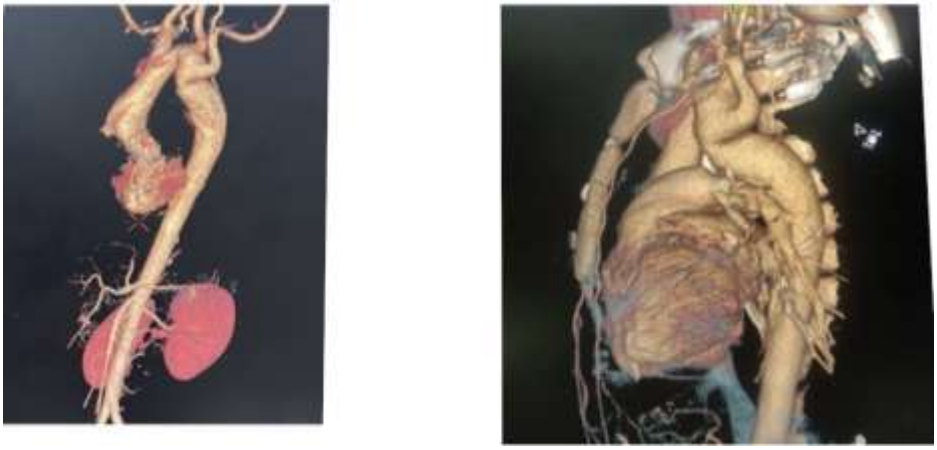

Figure 6:- 15-month-old male child with chest complaints. Volume-rendered reconstructed CT image depicting pseudo-coarctation of the aorta, characterized by elongation and folding of the aorta on itself, resulting in the appearance of coarctation.

Case 7:

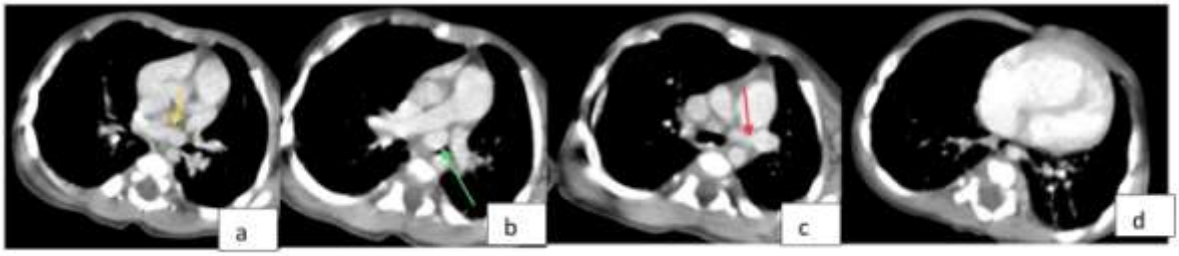

Figure 7:- 2-month-old male child with excessive crying and cyanosis. Axial post-contrast cardiac CT images illustrating: a) Formation of the confluence of the left upper and lower veins (indicated by the yellow arrow in a) into a vertical vein. b) The vertical vein (highlighted by the green arrow in b). c) Drainage of the vertical vein into the innominate vein (indicated by the red arrow in c), which subsequently drains into the right atrium. d) Enlargement of the right atrium.

Case 8:

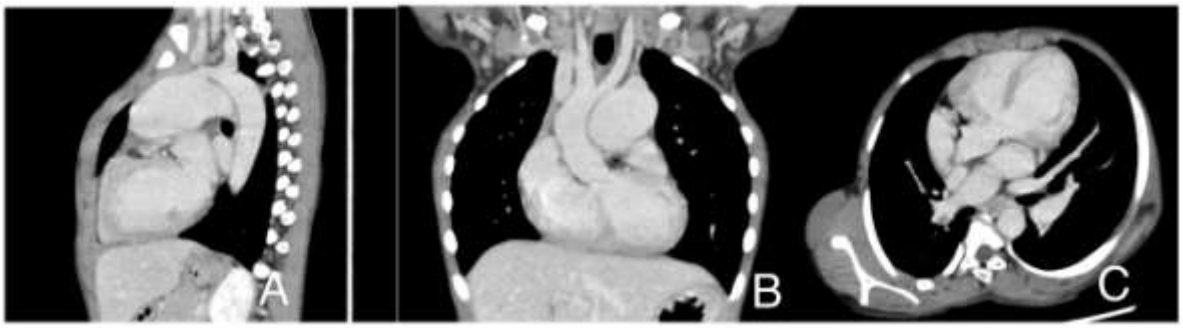

Figure 8:- 8-year-old male child with chest complaints with interrupted arch.A: Patent ductus arteriosus connecting the pulmonary trunk and the arch of the aorta. B: Termination of the proximal aorta into bilateral carotid branches, with the descending aorta and its branches receiving supply through the patent ductus arteriosus. C: Ventricular septal defect observed.

Case 9:

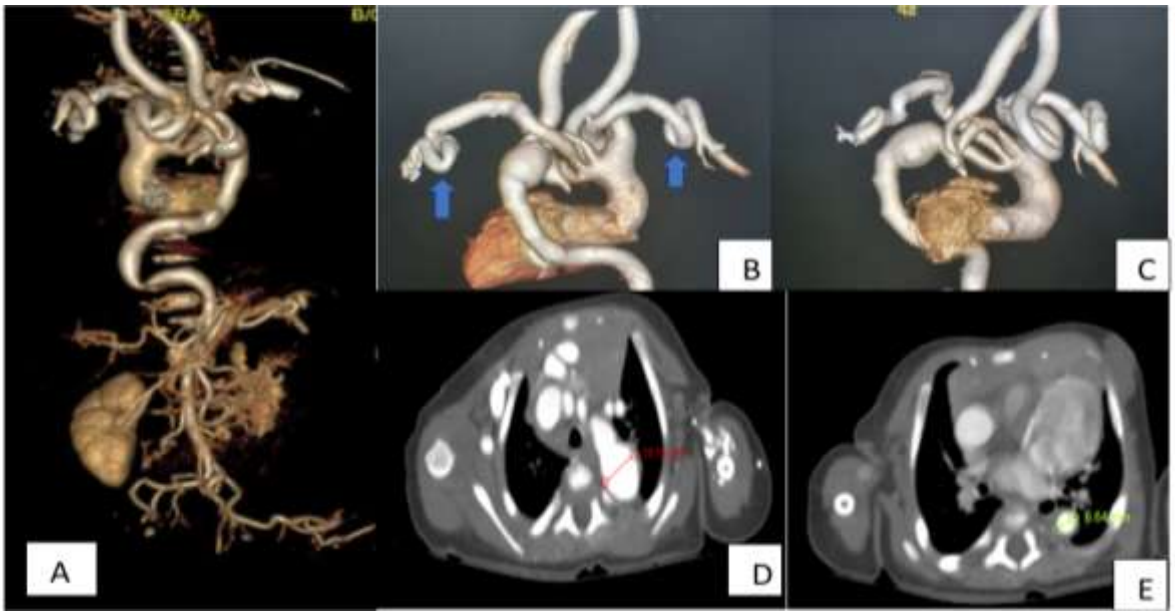

Figure 9:- 26-day old female child presented with complaints of swelling in left inguinal region and noisy breathing. Depicting CT images showing A. Volume rendered image depicting the entire aorta and its branches, with

elongation and tortuosity of most of the branches. B and C showing reconstructed images showing bilateral subclavian, carotids and their branches. D and E are axial post contrast CT images showing the calibre of the aorta and various levels.

Case 10:

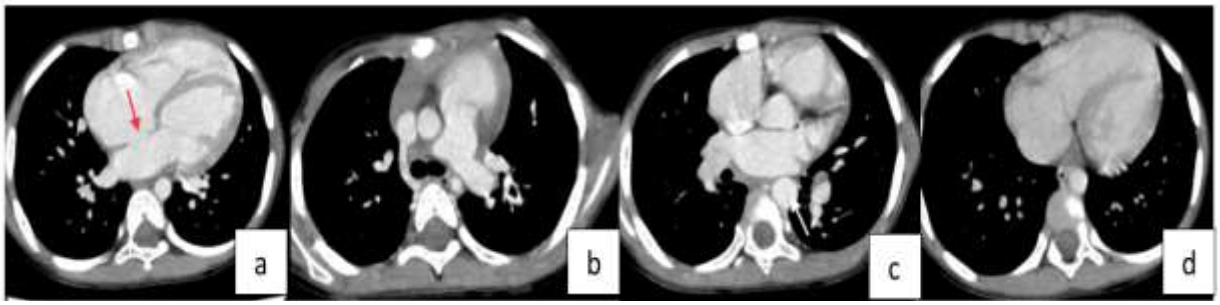

Figure 10:- 4-year-old female with weakness and tingling sensations. Axial post-contrast cardiac CT images illustrating: a) Atrial Septal Defect (ASD). b) Descending aorta at the level of coarctation, showing the narrowest lumen. c) Dilatation of a short segment of the aorta post-coarctation. d) Normal lumen of the descending aorta beyond the dilatation.

Case 11:

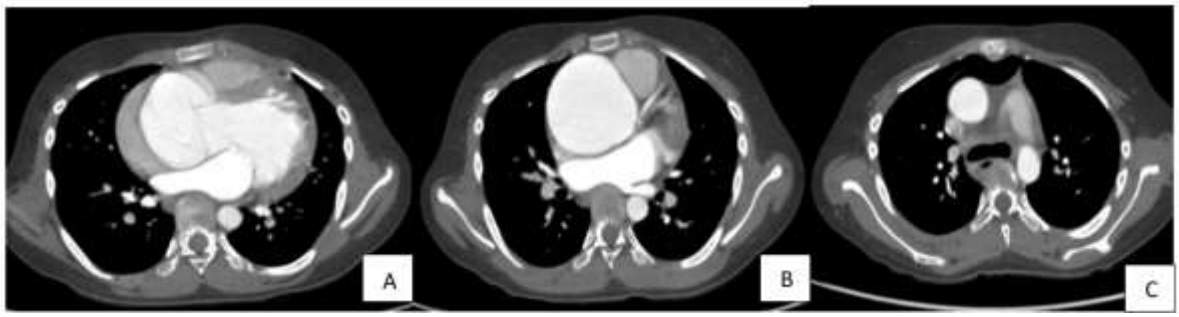

Figure 11:- 15-year-old with chest pain and breathlessness, diagnosed as fusiform aneurysm of ascending aorta. Depicted CT images showing A. Axial post-contrast image at the level of the aortic valve, revealing a dilated aortic root. B. Axial image displaying fusiform aneurysmal dilatation of the ascending aorta. C. Axial image of the same patient depicting normal calibres of the ascending and descending aorta at a level above the aneurysm.

Case 12:

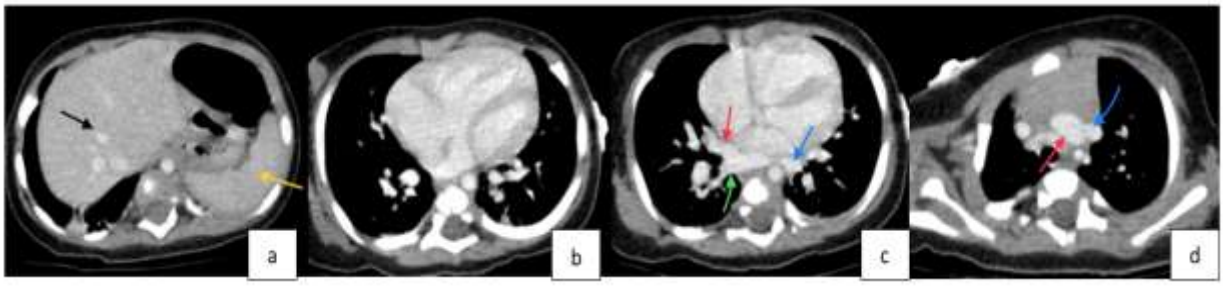

Figure 12:- 3-month-old who presented to the clinic with cyanosis. Axial post-contrast cardiac CT images illustrating: a) Normal abdominal situs with the liver on the right side (black arrow) and spleen on the left side (yellow arrow). b) D-loop of ventricles. c) Right upper pulmonary vein (red arrow), right lower pulmonary vein (green arrow), and left lower pulmonary vein (blue arrow) draining into the left atrium. Note the indentation caused on the left lower pulmonary vein at its junction with the right-sided veins by the descending aorta. d) Left upper pulmonary vein (blue arrow) draining into the innominate vein, indicative of left supra-cardiac Partial Anomalous Pulmonary Venous Return (PAPVR).

Case 13:

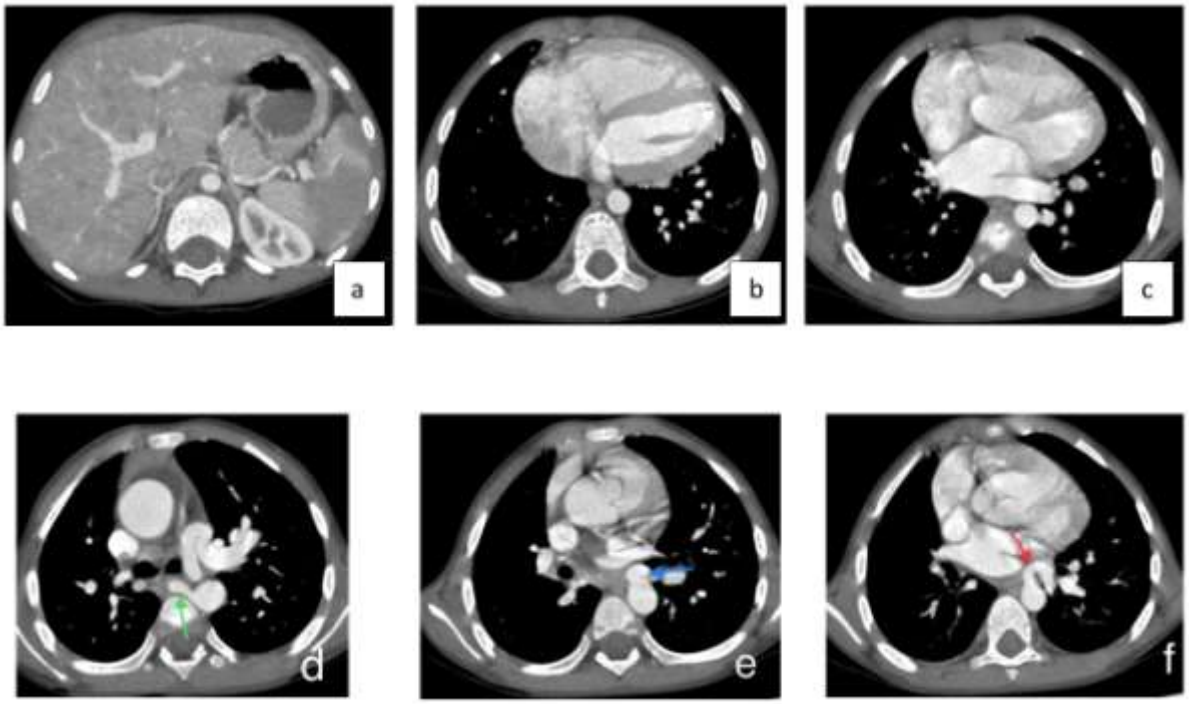

Figure 13:- 5-year-old who presented with complaints of shortness of breath and had reduced growth.Serial axial post-contrast cardiac CT images in a case of pulmonary atresia illustrating: a) Normal abdominal situs. b) D-loop of ventricles. c) Normal drainage of pulmonary veins into the left atrium. d) Major Aortopulmonary Collateral Artery (MAPCA) arising from the descending aorta supplying the right lung (green arrow). e and f) MAPCAs from the descending aorta supplying the left lung (blue and red arrows).

Case 14:

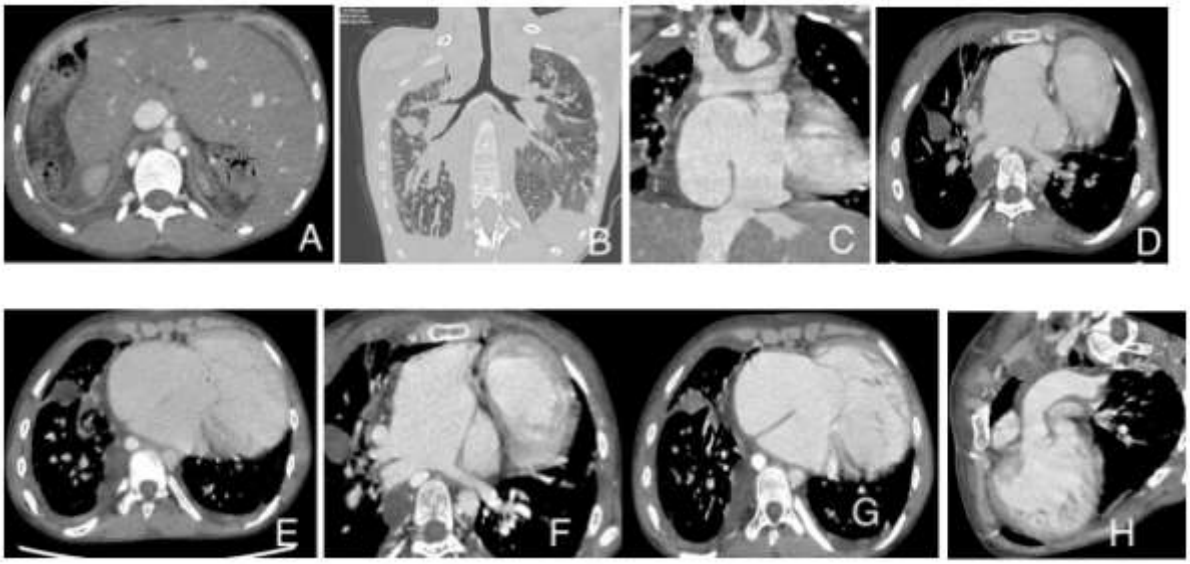

Figure 14:- CT images of a case of heterotaxy with right isomerism showing A: Midline liver predominantly in the left half and absent spleen with altered relation between abdominal aorta and inferior vena cava (IVC). B: Bilateral morphological right bronchus, suggestive of right isomerism. C: Bilateral superior vena cava (SVC). D: Levocardia and levoposition of the heart. E: Large ostium primum atrial septal defect (ASD), large ventricular septal defect (VSD), and atrioventricular (AV) valve defect. F: Pulmonary veins draining into the morphological right atrium located on the right side. G: Left atrium with an appendage similar in morphology to the right atrial appendage,

suggesting bilateral morphological right atria. H: Both outlets arising from the right ventricle, indicative of Double Outlet Right Ventricle (DORV).

Case 15:

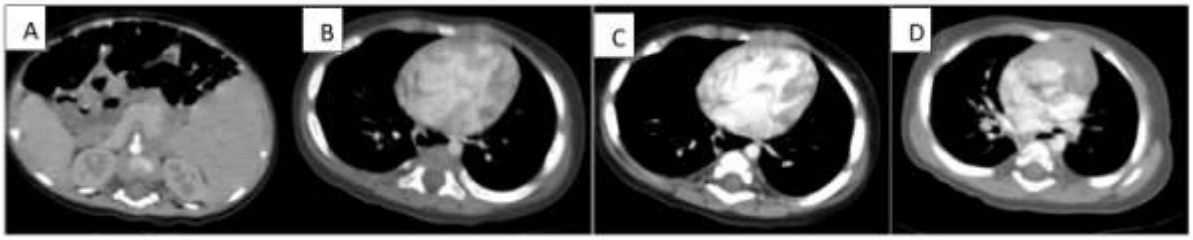

Figure 15:- 8-month-old with situs ambiguous, TGV, DORV common AV septal defect. Axial CT images depicting - A: Midline liver predominantly in the left half with the head of the pancreas in the left half and spleen in the right half, suggestive of situs ambiguous. B: Common atrioventricular (AV) septal defect and large ostium primum atrial septal defect (ASD). C: Hypertrophied right ventricle and hypoplastic left ventricle with a D-loop. D: Anteriorly placed aorta, valvular pulmonary stenosis, and Double Outlet Right Ventricle (DORV) (supravalvular and sub-valvular pulmonary stenosis also present, not shown in images).

Case 16:

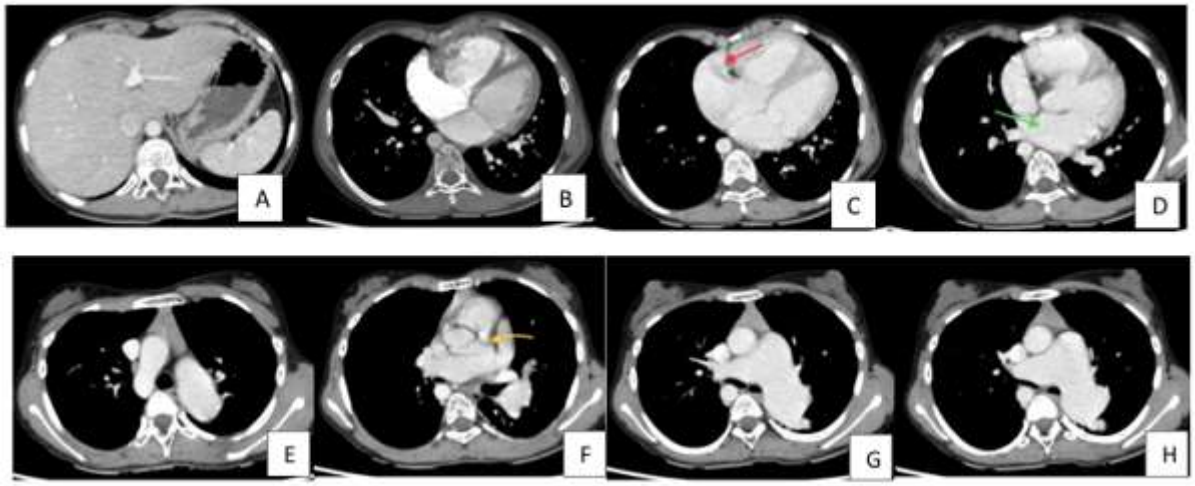

Figure 16:- 11-year-old case of tetralogy of fallot with right sided aortic arch. A- normal abdominal situs, B- D-looping of ventricles, C- morphological right atrium (right atrial appendage red arrow) on right side and left atrium on left side (green arrow) in D- suggestive of normal atrial situs. E – right aortic arch, F – pulmonary stenosis before bifurcation, G – right pulmonary artery and H- dilated left pulmonary artery.

Case 17:

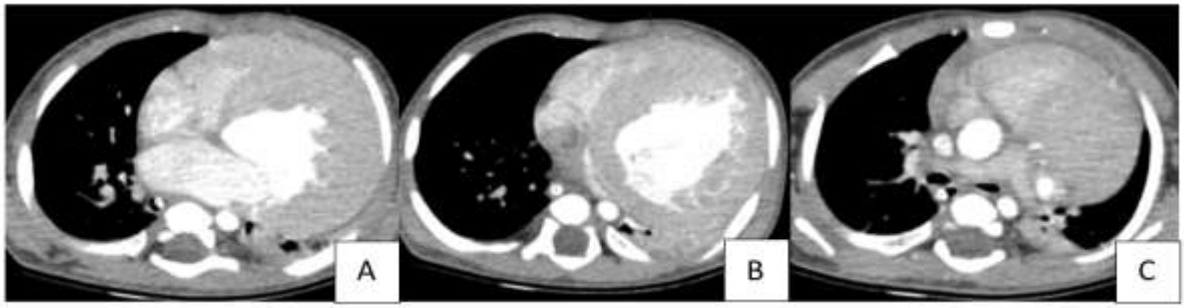

Figure 17:- Cardiac CT post contrast images of a case of left ventricular hypertrophy with pericardial effusion showing A: Left ventricular hypertrophy (LVH) and small right ventricle (RV). B: LVH and D-looping of the ventricles. C: Hypoplastic pulmonary arteries.

Case 18:

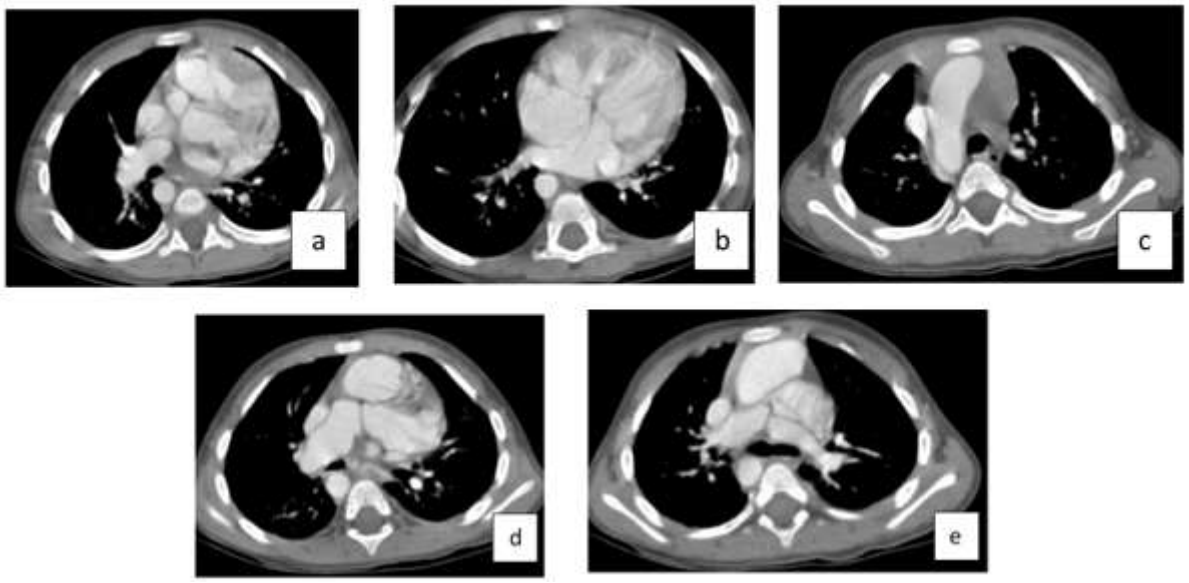

Figure 18:- 1 year old case of TOF**.** Axial post contrast cardiac CT images showing : a – anteriorly placed left ventricular outlet (aorta) to right ventricular outlet (suggesting altered relation between great vessels) and overriding of aorta, b – ASD and small VSD, c- right sided aortic arch (with mirror image branching pattern not shown in the image), d- dilated right pulmonary artery and e- left pulmonary artery(likely due to pulmonary stenosis at the level of infundibulum not shown in image causing post stenotic dilatation).

Case 19:

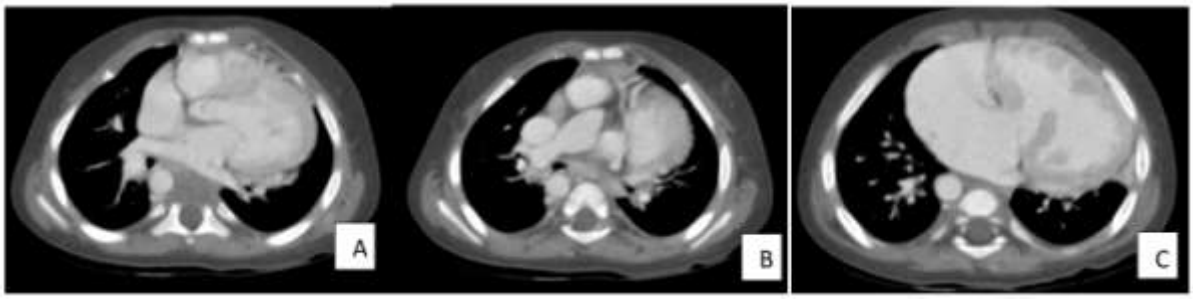

Figure 19:- 1 year old of transposition of great arteries and septal defects. Images depicting - A: Aorta arising from the right ventricle and pulmonary trunk arising from the left ventricle. B: Relation between great arteries, with the aorta positioned anterior to the pulmonary trunk, suggestive of D-Transposition of the Great Arteries (D-TGA). C: Large muscular ventricular septal defect (VSD).

Case 20:

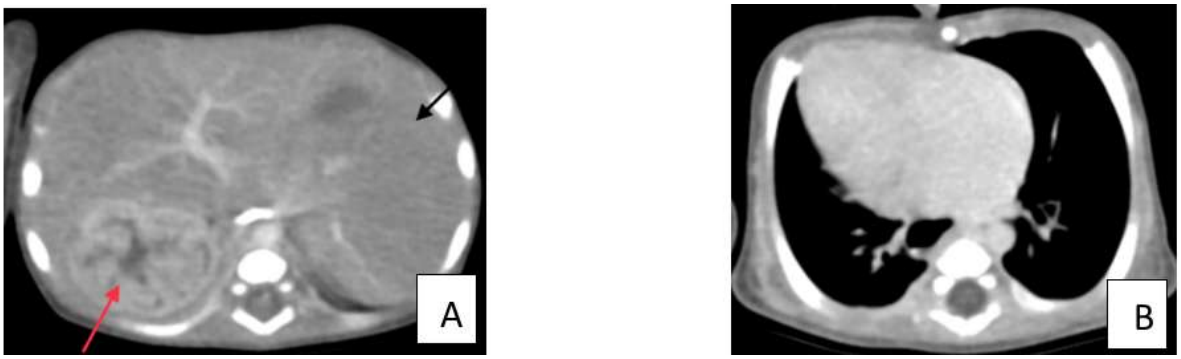

Figure 20:- 2-month-old male child with heterotaxy syndrome. Axial CT images illustrating: A: Liver (black arrow) midline in location predominantly occupying the left half, with the stomach (red arrow) on the right side and absent

spleen, suggestive of situs ambiguous. B: Dextrocardia, as the apex of the heart is located in the right hemithorax, indicating dextroposition of the heart.

Discussion:-

Key findings

The key findings from the cardiac CT scans align with previous research on congenital heart diseases, corroborating the notion of a heterogeneous spectrum of anatomical variations within affected populations [4]. For instance, the identification of atrial and ventricular septal defects, pulmonary atresia, and anomalies in aortic arches corroborates with established literature on common CHDs [1,5]. Moreover, the detection of less common anomalies such as interrupted arches, anomalous pulmonary venous returns, and ventricular hypertrophy is consistent with the understanding that CHDs manifest in diverse forms and presentations [6]. These findings collectively contribute to the body of knowledge surrounding CHDs and reinforce the need for comprehensive diagnostic approaches in clinical practice.

Review of Literature:-

The most common diseases projected to be present among CHD patients needing close observation include coarctation of aorta, aortic stenosis, tetralogy of fallot and ventricular septal defects [7].

In patients with CHD, some anatomical areas are inadequately characterised with trans-thoracic echocardiography, including the right ventricle, transverse and descending aortic arch, and pulmonary vasculature [8]. Although it often is useful for assessing these structures, trans-esophageal echocardiography has anatomical blind spots [9] and can cause airway compromise if the pulmonary artery is enlarged [10].

VSD is the most common congenital heart defect in the paediatric population and is second to atrial septal defect (ASD) in the adult population [11]. Tetralogy of fallot is classically described as a collection of four defects: infundibular or pulmonary stenosis, right ventricular hypertrophy, overriding aorta and VSD [12]. The incidence of bicuspid and congenitally dysplastic aortic valve is 1-2% in the general population [13]. Long term complications associated with bicuspid aortic valve include aortic stenosis, regurgitation, dissection, root dilation and endocarditis [14,15].

Strengths and limitations

The strengths of using cardiac CT scans to identify anatomical variations in congenital heart diseases lie in its ability to provide detailed, three-dimensional imaging of cardiac structures, allowing for comprehensive assessment of abnormalities. Additionally, cardiac CT is non-invasive and relatively quick, making it suitable for diagnosing CHDs in both paediatric and adult populations.

However, there are limitations to consider. Firstly, cardiac CT exposes patients to ionizing radiation, which can be a concern, especially for paediatric patients. Moreover, certain cardiac anomalies, particularly those involving complex vascular structures or dynamic blood flow patterns, may be better evaluated using other imaging modalities such as cardiac MRI or echocardiography. Additionally, interpretation of cardiac CT findings requires expertise in cardiac imaging and may be subject to variability among radiologists.

When interpreting results from cardiac CT scans, readers should consider the clinical context, patient demographics, and potential limitations of the imaging technique. Collaboration among multidisciplinary teams, including cardiologists, radiologists, and cardiothoracic surgeons, is essential for accurate diagnosis and treatment planning in patients with congenital heart

Conclusion:-

In summary, the study's analysis of cardiac CT scans in congenital heart disease patients uncovers a wide array of anatomical complexities, emphasizing the intricate nature of these conditions. It significantly contributes to our understanding by providing comprehensive insights into structural abnormalities, aiding in precise diagnosis and treatment planning. Achieving its objectives, the study identifies and characterizes various anomalies, enriching existing knowledge in this domain. This underscores the need for ongoing research and collaboration to refine diagnostic and management strategies, ultimately improving patient outcomes. While cardiac CT imaging proves

invaluable for its non-invasive nature and detailed anatomical information, its judicious use alongside other modalities and clinical data is essential, acknowledging both its benefits and limitations in optimizing patient care.

Ethical Considerations:

This study adheres to the principles outlined in the Declaration of Helsinki and is conducted in accordance with institutional research ethics guidelines. Patient confidentiality and privacy are strictly maintained throughout the study, with all data anonymized and stored securely. Institutional review board approval is obtained prior to commencement, and informed consent requirements are waived given the retrospective nature of the study. Measures are in place to mitigate potential biases and conflicts of interest, ensuring the integrity and validity of the research findings.

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