

BUNDLE OF DELIMAS IN DEXTROCARDIA AND AN OVERVIEW.

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Running Title: Orientations in Dextrocardia with ACS

Abstract

Dextrocardia is a rare non-fatal congenital anomaly. Most cases were diagnosed incidentally and throws lot of diagnostic and therapeutic challenges in general and management in particular. We come across 3 different cases with Dextrocardia. First case was asymptomatic young female for regular health check. Second case was middle aged male with Acute MI. Coronary Angiogram revealed lesion in RCA. Managed with Primary PTCA and Stent to RCA. Third case was elderly female with multiple comorbidities came with Positive TMT. Coronary Angiogram revealed TVD. Underwent CABG with 2 grafts. All three cases were treated successfully. We faced lot of Diagnostic to Therapeutic challenges. This includes - ECG & Chest Ray Interpretations, 2D Echo observations, Accessing and engaging coronary ostium, Contrast overload and Recognition of parts of cardiac surface anatomy especially during cardiac surgery. Dextrocardia is not an uncommon congenital cardiac anomaly. Its importance increases whenever it is associated with other cardiac illness in general and ACS in particular. Difficulty exists in both structural and functional assessment of the heart and coronary arteries. Various techniques are used to overcome these problems. Cautious and timely intervention is required. This will not only reduce the time taken for intervention (if required), but also minimizes the complications in a safe and cost-effective manner.

Key Words:

- ACS = Acute Coronary Syndrome
- ECG = Electrocardiogram
- TMT = Tread Mill Test
- ACS = Acute Coronary Syndrome
- IPWMI = Infero Posterior Wall Myocardial Infarction
- SVD = Single Vessel Disease
- PTCA = Per Cutaneous Coronary Angioplasty
- RCA = Right Coronary Artery
- TVD = Triple Vessel Disease
- CABG = Coronary Artery Bypass Graft

Text

Introduction: Heart has complex embryological development, includes both general growth and looping. Normal looping will result in **Levocardia** (Heart on left side - Normal) and Failure to Loop results in **Dextrocardia** (Heart on right side - Abnormal). Majority of these patients are asymptomatic, incidental finding and may be associated with other congenital anomalies too. On the contrary, Acute Coronary Syndrome (ACS) is an acquired and may be

emergency clinical situation. Occurs due to Significant Coronary Artery Disease (CAD), managed with either Medical Management or Revascularization, either by Catheter based Interventions or Surgical Coronary Revascularization. There are many Diagnostic and Therapeutic challenges in Interpretation of Cardiac Investigation and Management, demands better understanding of basic knowledge in general and orientations in particular. We have come across 3 different cases of Dextrocardia with IHD.

Presentations

Case 1 - Young Thirty-two-year-old female with no comorbid conditions and not on any treatment. Came to our hospital for regular General Health Check for the first time. She was asymptomatic with stable vitals (Afebrile, PR - 86/min, BP - 132/82 mm of Hg and SpO₂ - 95% @ room air). She underwent few investigations as per MHC Package, including ECG, Chest X Ray and 2D Echo. All investigations are within normal limits. Her Dextrocardia status was diagnosed incidentally and evaluated with ECG, CXR and 2D Echocardiography. She was counselled and managed conservatively.

Case 2 - Middle-aged Forty-Eight-year-old man with known Dextrocardia and on regular treatment for Hypertension with ACE inhibitor, was admitted with history of Acute chest pain for 4 hours duration, non-radiating and associated with sweating and generalised weakness. On admission - His vital signs are stable (Afebrile, PR - 74/min, BP - 112/74 mm of Hg and SpO₂ - 95% @ room air). His ECG suggestive of Acute Infero Posterior wall Myocardial Infarction. His 2D Echo was suggestive of Regional Wall motion abnormalities with LVEF of 45%. Immediately received Oral Antiplatelets, Statins, Anticoagulants and IV Fluid. Meanwhile relatives were counselled and consented for Primary PCI. After high-risk consent, immediately taken up for Coronary Angiogram (CAG), done through Left Radial Artery approach which showed SVD of RCA and without any Coronary Anomalies. Being a known Dextrocardia, as per our anticipation we had minimum difficulty in engaging the coronary ostium. Then successful Primary PTCA + Stent to RCA was done with 3.5 x 18 mm of Yukon Choice Flex Stent. Over all, nearly 20 ml of extra contrast load was required. Post PCI Serum Creatine was normal because he was adequately hydrated with IV Fluids prior to Primary PCI. Post procedure and his hospital stay was otherwise uneventful. His Dextrocardia status with ACS was evaluated and managed with ECG, CXR, 2D Echocardiography, Diagnostic Cath and RCA lesion Intervention.

Case 3 - An Elderly Seventy-Eight-year-old female with known Dextrocardia, but came without any previous records. She was on treatment for fairly controlled Type 2 DM and Hypothyroidism. This time she was presented with history of gradually worsened exertional chest discomfort from NYHA Class II to III since 1 ½ month. Her recent TMT for exertional ischemia, was reported as positive and hence she came for planned coronary evaluation and management. Basic investigations were done on Out Patient basis. On admission her vital signs are stable (Afebrile, BP - 132/78 mm of Hg, PR - 78/min, SpO₂ - 95% @ room air and GRBS - 208 mg/dl). On the same day, she underwent Coronary Angiogram which showed TVD. Advised CABG with grafts to LAD and RCA. Also advised medical management for Non dominant LCX lesion. Being a known Dextrocardia, we did not have any difficulty in engaging the coronary ostium. CTVS opinion was taken. Within next two days, she underwent preliminary and baseline investigations for CABG includes Bilateral Carotids,

USG Abdomen and Bilateral lower limb vascular doppler study. All the reports were within normal. On 4th day of admission, she underwent successful CABG (Coronary Artery Bypass Graft) with 2 grafts (SVG to LAD & SVG to RCA). Being a right-handed operator, surgeon stands left side of the patient, when chest was opened. Being all lesions are proximal, there was no difficulty in identifying the lesion site. Surgery was done under General Anaesthesia with Off Pump setup and conduits for graft were obtained from Endoscopic Vein Harvesting from Left SVG (Saphenous Vein Graft). Post procedure and her hospital stay was uneventful. She was recovered gradually. Her Dextrocardia status was evaluated and managed with ECG, CXR, 2D Echocardiography, Diagnostic Cath and Cardiac OT procedure.

Discussion

Dextrocardia is rare developed from incomplete complex embryological sequence of events with varied morphology.¹ Its Incidences and Events are summarized in **Table A** and **Table B**.² Different terminologies were used to define the Cardiac Position as in **Table C** and Few congenital associated anomalies were noted as in **Table D**.¹ The incidence of ACS with or without Dextrocardia is same, but may have variable symptoms.³ Chest pain may be from Right side, due to Visceral pain localized from Peripheral Nerve Root Transposition (50% in Situs Inversus).³ ACS demands quick and accurate assessment. The combination of IHD and Dextrocardia, needs special attention as this may cause variety of problems ranging from Diagnostic Delimas to Therapeutic difficulties. This is especially occurred when the patient presented as ACS with Unknown Dextrocardia status and Acute MI with Known Dextrocardia. Only counselling and assurance is enough for Dextrocardia only presentation. But in Dextrocardia with Significant CAD requires revascularization either by Cath Interventions or Bypass Surgery. Some special technique or hardware will be used, mainly to engage Coronary Ostium during Cath Diagnostics to Interventions as in **Table E**. We are here summarizing all our observations and standard protocols used on case-to-case basis.

Case 1 - Here the subject came for routine health check. ECG only can't be differentiated from Levocardia to Dextrocardia - if chest leads were placed respectively. While recording ECG, we initially thought that it is Technical Dextrocardia (**Fig 1A**) and ECG's Limb Leads were placed reverse inadvertently. But Cardiac Apex felt and sound best heard at Right mid clavicular line of Right 5th Intercostal Space. Hepatic dullness by Percussion present on right side confirms Situs Solitus. Suspect Situs Inversus if Hepatic dullness present on left side. Then regular ECG was taken by placing the chest leads in reverse fashion over the right side of the chest without changing the Limb leads. 2D Echo of TTE - from left side could not reveal anything and only Lung shadows were revealed. Confirm position of the Apex by Auscultation, then we place the probe over it. Then instead of turning the marker on left lateral side - we placed it on right lateral side to view the Apical cluster of 2D Echo Views. Then similarly PLAX and SAX views are obtained from right side, same level and opposite side turning of probe marker (**Fig 2A- Probe positions, Fig 2B & Fig 2C - TTE and TEE Images**). 2D Echo along with properly labelled Chest X ray confirms True Dextrocardia (**Fig 1B**). Discharged with counselling and assurances only.

Case 2 - Here the patient came with ACS - Acute IPWMI. He was already known Dextrocardia without any other problems. Being a known Dextrocardia, ECG recording and 2D Echo evaluation was done accordingly without any problem. On admission, his ECG was

initially taken from left side and then switched over to right side. Also, to avoid variations between serial ECGs, we make sure that chest leads should be placed at same place. This we have done by writing over the skin with a skin marker. This not only maintains the uniformity, but can also records any changes between ECGs. In view of Acute chest pain and ECG changes, relatives were counselled and consented for Primary PCI. During the procedure, there was little delay in engaging the Coronary ostia. This requires more than regular volume of contrast. With pre procedural IV fluids onflow, Contrast Induced Nephropathy (CIN) was avoided. With minimal volume of contrast, Aortogram and Aortic Focussed view was done to identify the level of Coronary Ostia and to engage it (**Fig 3A - Coronary Angiogram**). We also consciously ruled out any coexisted coronary anomalies. In this case also, 2D Echo (**Fig 2A- Probe positions, Fig 2B & Fig 2C - TTE and TEE Images**) and Chest X Ray (**Fig 1B**) were done. Noted to had similar delimas as in Case 1 and they were solved accordingly. Discharged with stable hemodynamic.

Case 3 - Here the patient came with Multiple comorbids and Exertional dyspnoea and TMT +ve. CAG reported as TVD for CABG. While performing CABG, after opening the chest surface anatomy of cardiac structures - which are identified in relation to chambers (**Fig 3B - Surgical Surface Anatomy**). And right-handed operator usually stands left side of the patient and viceversa. This is very important to identify the position of lesion and Graft. (**Fig 2A- Probe positions, Fig 2B & Fig 2C - TTE and TEE Images**). Here patient had Perioperative TEE during surgery. During TEE - the level will be same but only side changes. In this case, ECG, 2D Echo, Chest X Ray and Diagnostic Cath and On-table Surgical Delimas were continued and solved accordingly. Discharged with stable hemodynamic.

Conclusion

Dextrocardia is rare, benign and relatively asymptomatic cardiac congenital anomaly.² One must be careful to prevent wrong diagnosis or death due to delayed management,² especially with acquired conditions like ACS. One should have both Diagnostic and Therapeutic orientations in relation n to Dextrocardia. Diagnostic orientations need in ECG, Chest X Ray, 2D Echocardiography and Diagnostic Cath Studies. Whereas Therapeutic orientations need in Cath Interventions and during Cardiac Surgery. With this knowledge one can avoid wrong interpretations (of ECG, Chest X Ray and Echo), Prolonged Cath interventions (as fail to engage coronary ostia), Excess Radiation, Contrast overload (for Contrast Induced Nephropathy - CIN) and Wrong structural identifications during surgery. Major problem appears mainly if there is ACS with Unknown Dextrocardia and those who require immediate cath interventions - where time is precious. One should be cautious and aware of the fact that Dextrocardia may be associated with other congenital anomalies like Situs Inversus and Coronary Anomalies. May involve Multi-disciplinary approach and type of treatment varies according to different conditions and its severity.² Isolated Dextrocardia will lead normal life.³

Learning Points

- Levocardia with No Cardiac shadow seen in Morbid Obesity and Left Side Pneumothorax.
- In ACS with Dextrocardia:

- May require Multispeciality approach for diagnosis - Clinical examination to preliminary Investigations (ex. ECG), may have varied presentations.
- May presents with Right sided Chest pain
- Expect technical difficulties - Prior to Aortogram, reveals local coronary anatomy.
- Avoid procedural related complications
- Suspect Dextrocardia whenever there is:
 - Right sided Chest pain / Apex beat felt at Right Mid Clavicular Line of Right 5th Inter Costal Space / Recorded ECG shows chest leads reversal / Only Lung shadows or No Echogenicity by 2D Echo on left side
- Suspect:
 - Situs Inversus - Hepatic dullness on Right side by Percussion.
 - Coronary Anomalies - Difficult to engage coronary Ostium during Cath interventions.

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List of Tables

Table A: Sequence of events and Incidences:^{2,3}

Events and Incidences	
Incidence in Live births	1 in 12000
Incidence in General population	1 in 5000 to 30000
Male to Female Ratio	1 : 1
In relation to Viscera with Dextrocardia	

	Situs Solitus	32 - 35%
	Situs Inversus	35 - 39%
	Situs Ambiguous (Indeterminate / Isomerism)	26 - 28%
Incidence of other Congenital Cardiac Anomalies		
	With Dextrocardia + Situs Solitus	High
	With Dextrocardia + Situs Inversus	Low
Few first ones ...	1 st Cardiac Catheterization (CAG) done in	1973
	1 st CABG done in	1980
	1 st PCI done in	1987
	1 st Trans Radial PCI done in	2007
	1 st PCI in with Situs Inversus, by using multipurpose catheters	By Moreya et al

Table B: Brief summary of events during embryological development of Fetal Heart:²

Summary of events during Fetal Heart development	
•	Fetal Heart develops from Primitive Cardiac Tube
•	This may loops either to : Right side called as - Dextro Loop or D Loop and Left side called as - Levo Loop or L Loop
•	Morphologically
❖	Bulbous Cardis develops into - Right Ventricle
❖	Bulboventriculous develops into - Left Ventricle
•	In early stages of Fetal life, Cardiac Apex situated in Rt. Hemithorax
•	By the end of 1 st Month, Apex migrates to Lt. Hemithorax
•	Depending on the degree of migration, Apex position varies
•	It may be complete (Levocardia) or incomplete migration (Dextrocardia)

Table C: Features in relation to Heart Position:¹

Dextroposition	Dextroversion	Dextrocardia
RA, RV on Rt side of LA, LV	RA, RV on Rt side of LA , LV	LA on Rt. of RA
Major axis of heart pointing left	Major Axis of heart from left shoulder to right hip	Major axis of heart from left shoulder to right hip
Entire heart to right of midline / retrosternal	Apex to right of midline	Apex is in Right 5 th Intercostal space
Transducer to right, usual orientation of plane	PLAX plane in Mirror Image. PSAX normal orientation	PLAX and PSAX are in Mirror image
LA = Left Atrium : LV = Left Ventricle : RA = Right Atrium : RV = Right Ventricle : PLAX = Parasternal Long Axis View : PSAX = Parasternal Short Axis View		

Table D: Few Associations with Dextrocardia:¹

Situs Inversus	Internal organs of abdomen are positioned on the opposite side
Situs Solitus	Internal organs are not affected
Mesocardia	The heart is placed in the midline of thorax. Apex of the heart is either in the midline or directed towards right

Heterotaxy	This condition causes mirrored organs in chest and abdomen
Polysplenia Syndrome	Dextrocardia + Isomerism
Asplenia	Means born without a spleen a/w heterotaxy
Malrotation	People with intestines have not formed properly and they may coil in the wrong direction
Kartagener syndrome	This is a type of primary ciliary dyskinesia, caused by a genetic mutation - that affects the cilia in lungs leads to frequent infections

Table E: List of Special Techniques and Hardwares to engage Coronary Ostium:³

Double Inversion Technique (Horizontal Sweep - Reverse Display)	• Reverse the Image Acquisition
	• Horizontal Inversion of on-screen reversal of Images
Catheter Rotation (Torquing) Technique	• Counter-Clockwise Rotation is required to engage Left sided RCA with Right Judkins Catheter
Type of Catheter	• Left Judkins Catheter - used to engage Right sided morphological Left Coronary Artery ostium
	• Right Judkins Catheter - used to engage Left sided morphological Right Coronary Artery ostium
Downsizing of the Catheter	• Downsizing of the catheter from 6F to 5F

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Figure 1: ECG and Chest X Ray Orientations in Dextrocardia:

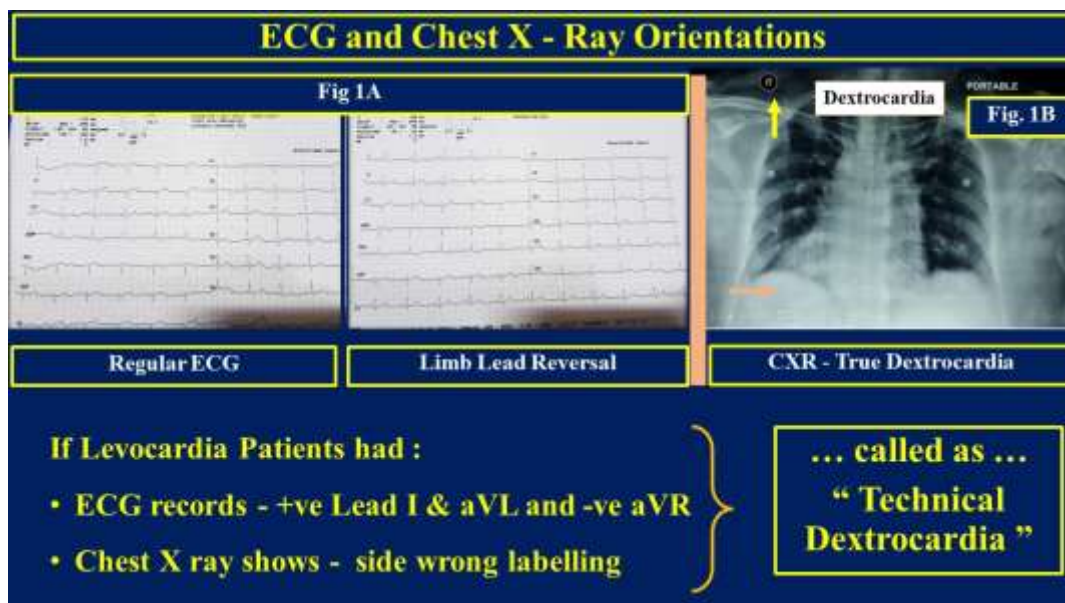


Figure 2: 2D Echocardiography Orientations in TTE and TEE:

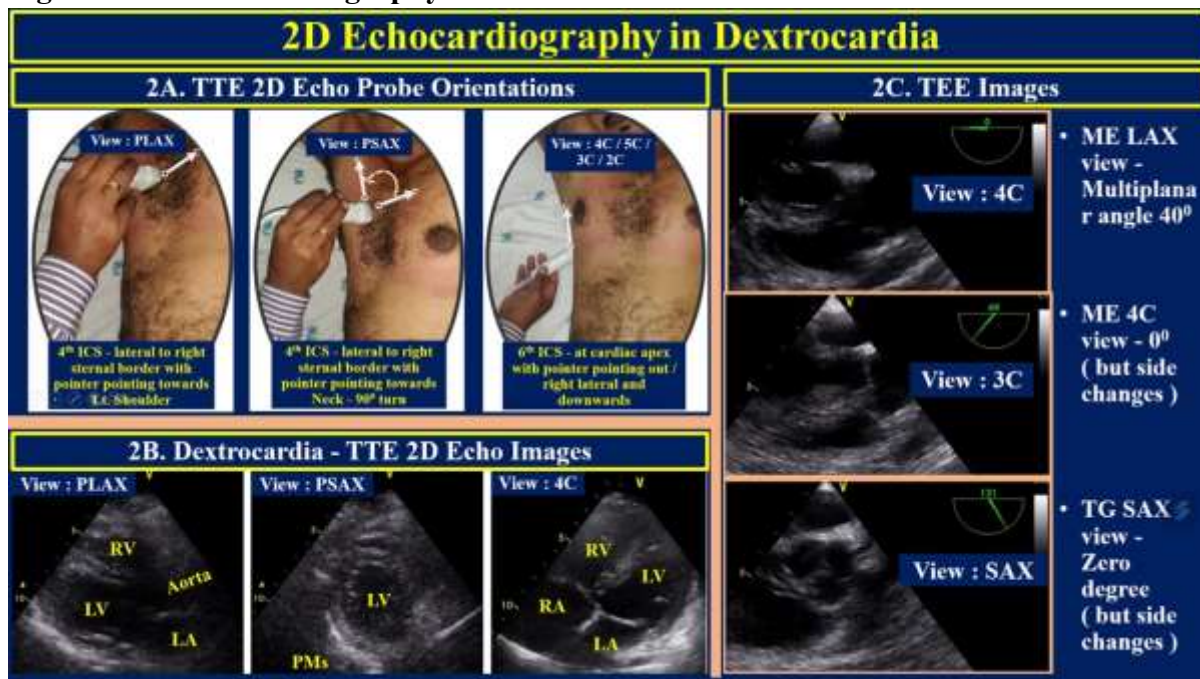


Figure 3: In Cathlab and On-table surgical Orientations in Dextrocardia:

