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Interpretation of 200 Children

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Abstract: *Evaluation of chest radiograph findings in 200 children\\\'s below 5 years over a period of 10 months. The aim of this study was to evaluate the findings of the chest radiographs after exposing the child *patients to the harmful radiation , no d11oubt: X-rays can be helpful, even life-saving but exposure to ionizing radiation causes damage to living tissue, and can result in mutation, radiation sickness and cancer. 0ne rare case with vertebral segmentation defects, ribs anomaly and associated dextrocardia which was presented on 20 days of her life with progressive respiratory insufficiency was detected. The neonate had multiple left lower ribs agenesis and bifid rib of right first and second ribs, dorsal left hemivertebrae in the mid and lower thoracic region with mild scoliosis of lateral convexity towards the right. Left hemidiaphragm is relatively elevated resulting in small thoracic volume and limited chest expansion; all consistent with a clinical diagnosis of Jarcho Levin syndrome with thoracic insufficiency. In addition, the neonate had Cardiac outline dearranged with apparent direction of cardiac apex towards the right side (Dextrocardia).Another rare case of cystic adenomatoid malformation of lung is noticed with respiratory insufficiency and one case with foreign body to the main bronchi causing complete collapse of left lung is recorded.*

Keywords*: Jarcho Levin syndrome, Dextocardia ,chest x rays.*

I. Introduction

Jarcho–Levin syndrome is an eponym that represents a spectrum of short-trunk skeletal dysplasias with variable involvement of the vertebrae and ribs. Other abnormalities have also been described in Jarcho–Levin syndrome, including neural tube defects, Arnold–Chiari malformation, renal/urinary tract abnormalities, hydrocephalus, hydroureteronephrosis, and meningomyelocoele [(1-3)]. Extensive deformities of the thoracic cage may affect the function and growth of lungs leading to its inability to support normal respiration. Jarcho Levin syndrome (JLS) is a rare lethal cause of such respiratory insufficiency. It is characterized by vertebral segmentation defects leading to congenital scoliosis and fusion of ribs. It was first reported by Jarcho and Levin in 1938.

0ver 120 cases of JLS are reported since 1938 and The ratio of reported spondylocostal dysplasia to spondylothoracic dysplasia is 1:6. Here is one such case of JLS with associated dextrocardia presenting in early neonatal period.

Interpretation of children's chest x rays after exposing to the harmful ionizing electromagnetic radiation (x rays).The interpretation of the child's chest radiograph should in theory be easier than the interpretation of the x rays of adult, in that the pathological entities that may be present are rather less numerous. However as we know in an infant it is impossible to control respiration and frequently ,movement. It is ,therefore, not easy to ensure that the films are taken in full inspiration in addition we are exposing the child to harmful radiation. Chest x rays can also reveal fluid in or around the lungs or air surrounding a lung however repeated advice of x rays to a child patient should be limited as the kids are still growing, they're more sensitive to radiation

II. Aims And Objectives

Interpretation of children's chest x rays after exposing to the harmful ionizing electromagnetic radiation (x rays).The interpretation of the child's chest radiograph should in theory be easier than the interpretation of the x rays of adult, in that the pathological entities that may be present are rather less numerous. However as we know in an infant it is impossible to control respiration and frequently ,movement. It is ,therefore, not easy to ensure that the films are taken in full inspiration in addition we are exposing the child to harmful radiation. Chest x rays can also reveal fluid in or around the lungs or air surrounding a lung however repeated advice of x rays to a child patient should be limited as the kids are still growing, they're more sensitive to radiation.

III. Materials And Methods

Source of data- All paediatrics patients referred for chest x ray, age below 5 years for a period of 8 months (March 2015-Oct 2015) Inclusion criteria -Both female and male child below 5 years. Exclusion criteria -children's above 5 years.

IV. Results

Table 1-Total number of pts

SEX	TOTAL	PERCENTAGE
Male	109	54.5%
Female	91	45.5%
Total	200	

Table 2-Chest Radiograph Findings

X ray Diagnosis	No of cases	Percentage
Vertibral anomely with normal chest findings	1	0.5%
Vertbral and rib anomely with dextrocardia	1	0.5%
Pleural effusion with consolidation	8	4%
Only pleural effusion	8	4%
Mediastinal lymphadenopathy	6	3%
Dextrocardia	2	1%
Cavitory lesions including one CCM of right lung.	8	4%
Cardiomegaly	4	2%
Consolidation of lungs	19	9.5%
Foreign body in left main bronchus	1	0.5%
Normal chest	142	71%

Table 3-Age wise distribution

Age of child	Numbers of pts who have taken x ray
1year	14%
2 year	11%
3 year	24%
4 year	21%
5 year	30%
Total number of pts	200

Out of 200 pts, a rare case found was of a 2.8 kg female baby Jessica, born at 34 weeks pregnancy, a second baby for her mother out of a non-consanguineous marriage presented on 20 days of her life in our department for taking a chest x ray with severe respiratory distress and the patient was admitted to our hospital on dated 6.8.2015 at 6.32 PM with MRD N0-15160006664 and was discharge on 15.8.2015. Baby had short trunk with scoliosis to the right. There was no history of similar bony defects in any member of the family. Her weight and head circumference were 2.8 kg and 31 cm, both corresponding to 50^{th} centile while the length of 31 cm was less than 3^{rd} centile. Upper to lower segment ratio was 1.3:1 (normal is 1.7:1) suggestive of short trunk dwarfism. Dextrocardia with congenital multiple ribs agenesis on left side with bifid right first and second ribs and hemivertibrae on left side [1]. Ultrasound of the abdomen and cranium was done by me and come out to be normal. Baby succumbed due to progressive respiratory failure over next 2 days. A clinical diagnosis of Jarcho Levin syndrome (JLS) with lethal progressive respiratory insufficiency with dextrocardia was made .Another case of foreign body to left main bronchus with complete collapse of left lung was also seen which was confirmed after performing HRCT and also succesfully removed in ENT OT. One more rare case of conginital cystic adenomatoid malformation of lung was recorded involving on right lung..Other findings of the study like consolidation of lung parenchyma, pleural effusion,cavitory lesions, mediastinal lymphadenopathy are also recorded commonly in our department as given (Table-2).

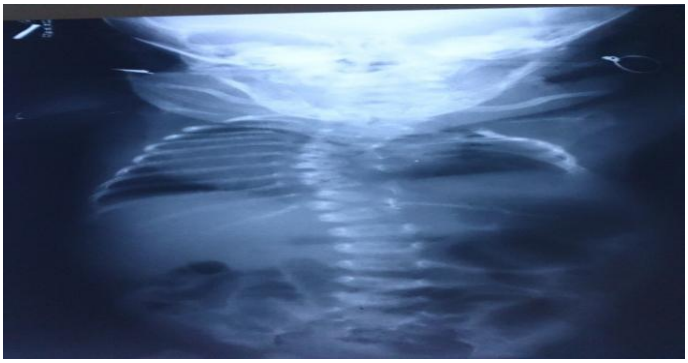


Fig 1 -X Ray PA view- Multiple left lower ribs agenesis and bifid rib on right side, dorsal left hemivertebrae in the mid and lower thoracic region with mild scoliosis of lateral convexity towards the right with dextrocardia. Left hemidiaphragm is relatively elevated resulting in small thoracic volume and limited chest expansion
g 2 -x ray pa view of a child of 2 yrs old with -Hemivertebrae on left side.

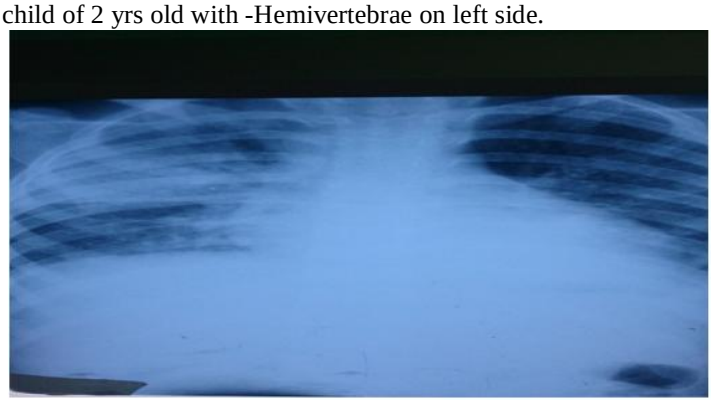
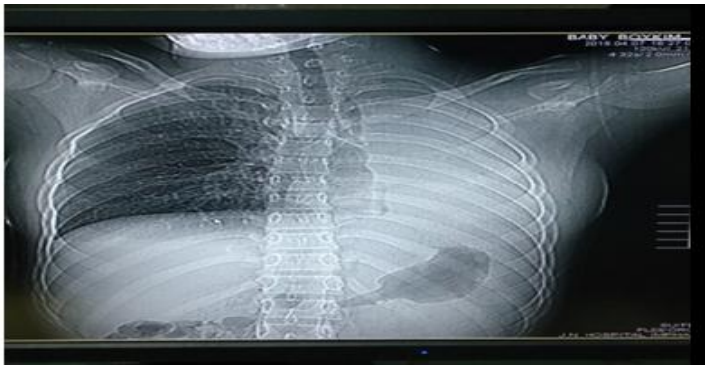


Fig 3 -X Ray PA view of a 5 yrs old child with Cardiomegaly due to conginital heart defect.



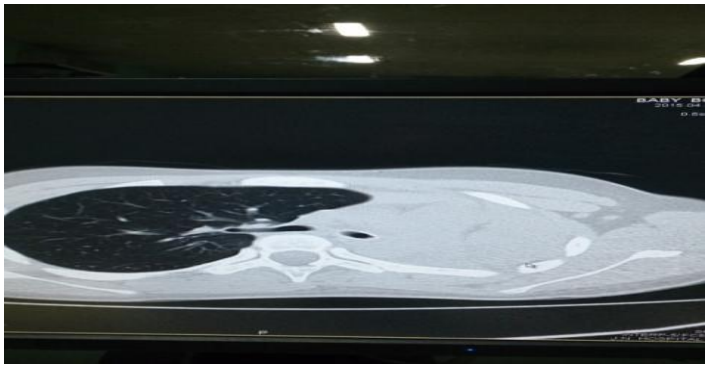


Fig 4A -X Ray PA view -Collapse of left lung due to foreign body at the left main bronchus. Fig 4b showing HRCT image of axial cut showing obliteration of left main bronchi resulting collapse of left lung with compensatory emphysematous right lung and shifting of anterior junctional line on the left side.

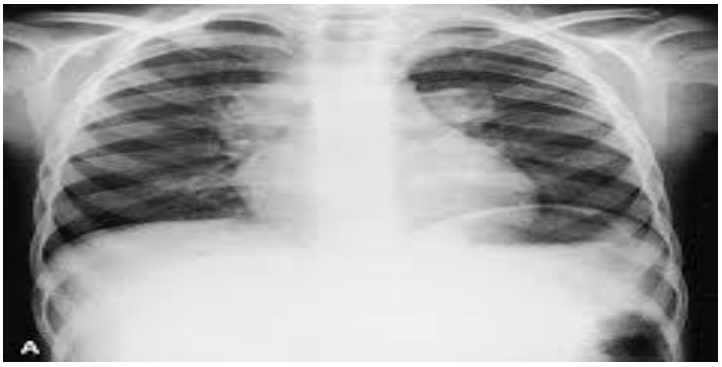


Fig 5- A bilateral hilar and mediastinal lymphadenopathy in a young boy of 5. This is case of primary infection.

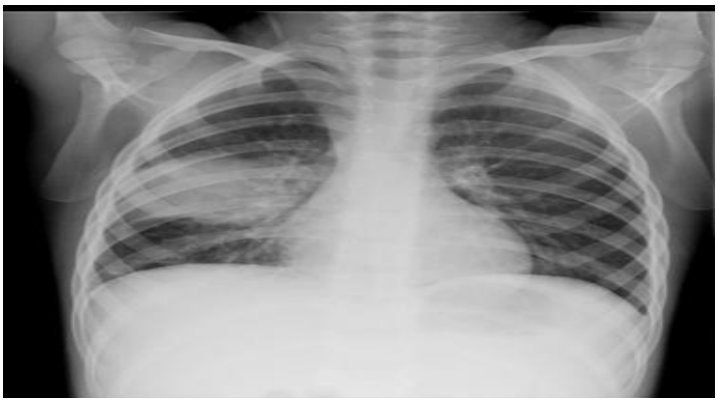


Fig 6 -Consolidation in the right mid zone of lung in a child of 4 years.

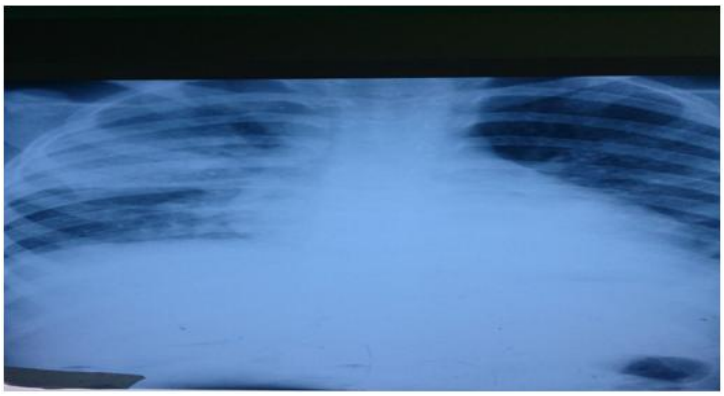


Fig 7- Well defined rounded consolidation of right upper lobe which on HRCT confirmed to be a cavitory lesion with surrounding consolidation and associated cardiomegaly.

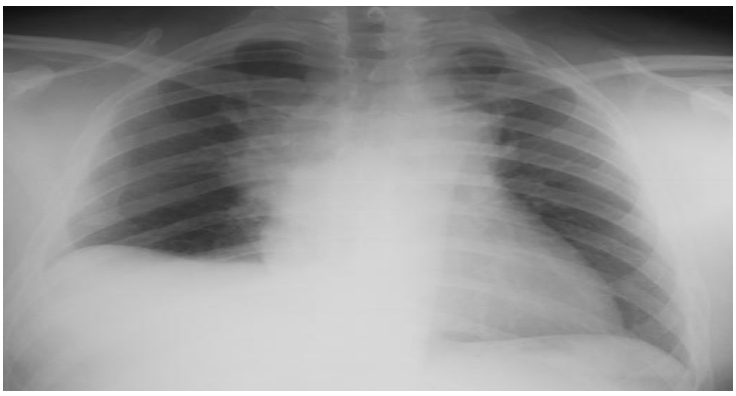


Fig 8- Right sided pleural effusion which is located in sub pulmonic area beneath the lung which was confirmed by HRCT

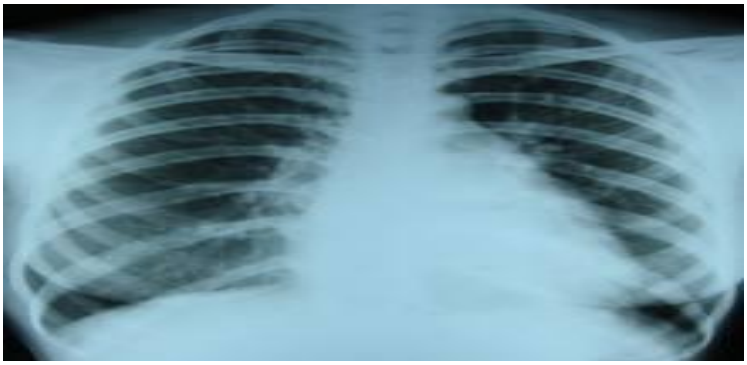


Fig 9- Mild enlargement of left atrial appendage with straightening of left heart border due to mitrial stenosis in a 5 years old female child.

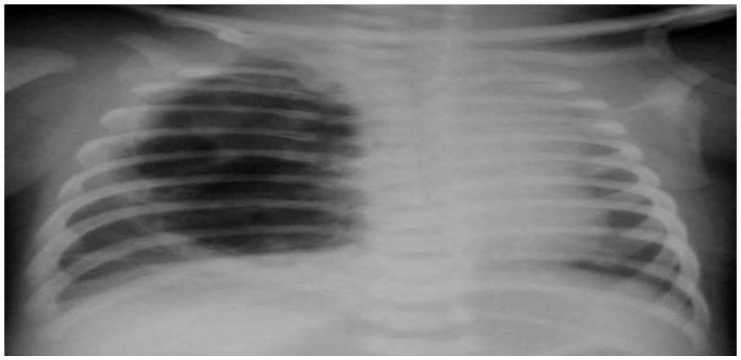


Figure 10. 4 and half year old boy presenting with irregular cystic lesions with associated soft-tissue elements in the right thorax, which had displaced the mediastinum to the left side. CT confirmed that it is a case of Cystic adenomatoid malformation of lung.

V. Discussion

Thoracic skeletal anomalies present as a rare cause of progressive respiratory insufficiency. Extensive vertebral and rib anomalies like Jarcho Levin syndrome (JLS), Juene syndrome and Ellis Van Crevald syndrome affect thoracic function and growth, which in turn affects lung growth. Shortening of the thoracic cage due to these anomalies leads to thoracic insufficiency.[4] Our case was fitting into the diagnosis of a JLS with thoracic insufficiency due to the presence of multiple hemivertibrae, multiple ribs agenesis ,bifid rib and scoliosis with limited thoracic volume and expansion leading to progressive respiratory failure.

JLS was first reported in 1938 in Puerto Rican origins by Jarcho and Levin.[5] They described a syndrome with a spectrum of vertebral segmentation defects and rib anomalies. Later in an article in 1996 Mortier *et al*. classified congenital vertebral segmentation defects into three subtypes - Jarcho Levin syndrome (JLS), spondylothoracic dysostosis (STD) and spondylocostal dysostosis (SCD).[6] Individuals with spondylothoracic dysostosis (STD) have an autosomal dominant inheritance pattern. Vertebral segment anomalies in spondylothoracic dysostosis (STD) spare the sacrococcygeal region and they do not have intrinsic rib anomalies. Spondylocostal (SCD) is a benign form and follows autosomal recessive inheritance. It is characterized by involvement of > 10 contiguous vertebral segments, pebble beach appearance of vertebrae on X-ray, involvement of sacrococcygeal region with intrinsic rib anomalies.[7,8] Jarcho Levin syndrome (JLS) is a lethal subtype which presents with vertebral segmentation defects like hemivertebrae or block vertebrae throughout the spine with fusion of ribs at cost vertebral junction bilateral asymmetrically leading to crab like appearance on X-ray. There is the presence of scoliosis and absence of ribs without intrinsic rib anomalies such as bifid, broadened or fused ribs. Diagnosis is essentially clinical. They usually succumb to respiratory insufficiency due to restrictive lung disease. Usually death due to respiratory insufficiency in the severe form of this condition commonly occurs within the first 2 years of life.[9]

Jarcho Levin syndrome (JLS)has been reported to be associated with cardiac, urogenital, digital and neural tube defects.[10] In a review, Hatakeyama *et al*.[8] reported association of congenital heart disease in 9 out of 87 cases of JLS. In 1976, Elier JL and Morton JM first reported a case in which diastematomyelia occurred in association with findings characteristic of the Jarcho–Levin syndrome in an infant born to a woman who abused lysergic acid diethylamide during pregnancy.[11] Malformations of the spinal cord with a separation into two hemicords have been termed "diastematomyelia." The term "diplomyelia" is reserved as a true duplication of the spinal cord which is, however, difficult to demonstrate. In 1992, Pang *et al.* recommended the term "split cord malformation" for all double spinal cords. A type I split cord malformation consists of two hemicords, each contained within its own dural tube and separated by a dura-sheathed rigid osseocartilaginous median septum. A type II split cord malformation consists of two hemicords housed in a single dural tube separated by a nonrigid, fibrous median septum.[12]

The Vertical Expandable Prosthetic Titanium Rib (VEPTR) was approved by the FDA in 2004 as a treatment for thoracic insufficiency syndrome in pediatric patients. Vertical Expandable Prosthetic Titanium Rib (VEPTR) is a device that helps straighten the spines and seperate ribs so that the lungs can grow and fill with enough air to breathe.The lenght of the device can be adjusted as the child grows.

VI. Conclusion

Chest radiograph is accessible as a potent diagnostic tool throughout the country when the cost benefit is evaluated. Chest x ray remains a major diagnostic modality, within the reach of common man in spite of its hazardous electromagnetic radiation which if exposed to often enough, has been linked to a greater chance of

getting cancer later in life however it should be the first choice of investigation before sending a patient to CT chest which again will exposed to more higher radiation.

Effective radiation dose according to **Grainger & Allison's**
Chest x ray - **0.02** mSv
CT Chest - **8** .**8** mSv
1 Ct Chest = **400** chest x- rays.

"No patients should be exposed to more radiation than they need at any age," says pediatric radiologist Marta Hernanz-Schulman, MD, chair of the American College of Radiology's Pediatric Imaging Commission.

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