INTRODUCTION

Chest wall malformations (CWMs) represent a wide spectrum of anomalies, with a relatively high incidence and a significant impact on the life of patients. Besides a minority of cases with functional respiratory impairment and symptoms, the clinical importance of these anomalies derives primarily from the fact that the majority of children and their parents seek medical advice for psychosocial concerns, sometimes severe, usually due topoor cosmesis and aversion to sports and public exposure. Despite the relatively high incidence, of CWMs are often misdiagnosed or neglected by physicians, thus resulting in asignificant delay or mistakes in the diagnostic work up or in the therapeutic management. Inthe last 12 years, however, since the introduction of the Nuss technique for pectusexcavatum (PE) (Nuss et al., 1998) the interest of the scientific community about CWMs has dramatically increased, as well as the number of publications on this topic. A wide range of CWMs exist. Some malformations are very well defined and others are part of a wide spectrum of deformities. Confusion still exists in the literature about CWMs nomenclatureand classification. A classification is of paramount importance because of the treatment implications. Other controversial issues are the treatment options: many different surgicaltechniques or other therapeutic alternatives have been proposed, especially in the last decade, so it can be difficult for a pediatrician or even a surgeon to advise correctly the patients about the possible correction techniques. In this chapter we will propose a simple classification, published few years ago by Acastello (Acastello, 2006).

Examples of chest wall deformities:

Kyphoscoliosis

Kyphoscoliosis is a disease of the vertebral column and its articulations, characterized by the simultaneous presence of scoliosis (lateral curvature of the spine) and kyphosis (antero-posterior spinal curvature). Signs of the latter may be destruction of vertebral bodies, bone tuberculosis, osteoporosis, etc. Primary or idiopathic kyphoscoliosis is a hereditary disease characterized by defects in the development of the vertebrae or by connective tissue disease. It accounts for 80% of cases and is more frequent in women. Secondary kyphoscoliosis generally originates in childhood neuromuscular diseases such as poliomyelitis or muscular dystrophies ***(McCool, Rochester DF 2000).***

Ankylosing spondylitis

Ankylosing spondylitis is an inflammatory disease of the axial skeleton that mainly affects males. Ninety-five percent of affected individuals share the genetic marker HLA-B27. Inflammation compromises the ligamentous structures of the spinal column, the sacroiliac articulations, and the ribs, leading to the fibrosis and ossification of these structures and, thus, to an increase in rib cage stiffness due to ankylosis and fusion of the costovertebral and sternoclavicular articulations ***(Van Noor et al, 1991).***

Pectus excavatum

Pectus excavatum is the most common deformity of the anterior thorax. Consisting of a very notable concavity of the lower third of the sternum, pectus excavatum is a congenital defect which is manifested in different degrees of severity. Thus, it is classified according to the extent of concavity visible in a lateral x-ray of the thorax. It is considered significant when the distance between the surface of the anterior wall of the thorax and the deepest part of the depression is greater than 3 cm. Its etiology is unknown, although a close relation to the Marfan syndrome has been found to exist, suggesting that the connective tissue around the sternum is altered. Lung volumes are at the lower limits of normal or slightly diminished, and compliance is normal unless accompanied by severe kyphoscoliosis ***(Haller et al 1996).***

The diaphragm is the major respiratory muscle, contributing to 75% of resting lung ventilation, with an excursion of 1–2 cm. During forced breathing, its excursion reaches 7–11 cm, variable with individual characteristics and methods ***(Quigley et al 1996).***

Diaphragm mobility in chest deformities:

Diseases that constrict the skeletal structure of the chest wall and the spine with its articulations may interfere with the functional capacity of the diaphragmatic pump, facilitating the development of respiratory insufficiency and failure ***(Sahebjami h and gartside 1996).***

The evaluation of diaphragmatic mobility has been traditionally performed using fluoroscopy. Although this method is considered the gold standard, it has some limitations, such as visualization of the diaphragm through a single incidence, need for corrective calculations and exposure patient to ionizing radiation.

In recent years, ultrasound has also become used to evaluate diaphragmatic mobility it offers some advantages over fluoroscopy including the lack of ionizing radiation and the possibility of use at the bedside of the patient, and direct quantification of the movement of the diaphragm. So ultrasongraphy has been shown to be a promising tool in the evaluation of the diaphragm function ***(Sahebjami h and Gartside, 1996).***

AIM OF THE WORK

Assessment of the diaphragmatic mobility by chest ultrasound in patients with chest deformities (developmental or acquired) with correlation of the results with the severity of the disease.

HISTORY OF ULTRASOUND

The use of ultrasound in medicine began during and shortly after the 2nd World War in various centres around the world. The work of Dr. Karl Theodore Dussik in Austria in 1942 on transmission ultrasound investigation of the brain was the first published work on medical ultrasonics. From the mid-1960s onwards, the advent of commercially available systems allowed the wider dissemination of the art. Rapid technological advances in electronics and piezoelectric materials provided further improvements from bistable to greyscale images and from still images to real-time moving images. The technical advances at this time led to a rapid growth in the applications to which ultrasound could be put. The development of Doppler ultrasound had been progressing alongside the imaging technology but the fusing of the two technologies in duplex scanning and the subsequent development of colour Doppler imaging provided even more scope for investigating the circulation and blood supply to organs, tumours, etc. The advent of the microchip in the 1970s and subsequent exponential increases in processing power have allowed faster and more powerful systems incorporating digital beam forming, more enhancements of the signal and new ways of interpreting and displaying data, such as power Doppler and 3-dimensional imaging ***(Bolliger et al., 2009).***

Chest Ultrasonography Overview

Physics of ultrasonography:-

Diagnostic ultrasonography is the only clinical imaging technology currently in use that does not depend on electromagnetic radiation. This modality is based on the properties of sound waves, and hence the mechanical and acoustic properties of tissues. Diagnostic ultrasound is mechanical energy that causes alternating compression and rarefaction of the conducting medium, traveling in the body as a wave usually at frequencies of 2–10MHz. In general it is assumed that the speed of sound in tissue is constant at 1,540 m/s ***(Middleton et al., 2004).***

When a pulse of ultrasound energy is incident upon the body, it interacts with the tissue in a variety of ways. Some of the incident energy is directed back towards the source and is detected. The time delay between the energy going into the body and returning to the ultrasound probe determines the depth from which the signal arises, with longer times corresponding to greater depths. This information is used in the creation of an image. Other factors that make the tissues distinguishable on a screen are their slightly different acoustical properties; one is known as the acoustic impedance ***(Hedrick et al., 2004).***

At the boundary between two different tissue types the sound waves can be:- (a) Reflected, like light off a mirror, this being the primary interaction of interest for diagnostic ultrasound, as it allows the major organ outlines to be seen; the diaphragm and pericardium are specular reflectors; (b) Refracted, like light rays passing through a lens and hence having their directions altered; (c) Scattered, like sunlight in the sky, sending sound waves off in different directions; this occurs when the ultrasound wave encounters a surface that is ‘rough’ and (d) Attenuated or absorbed, as they lose energy, which is converted to heat in the tissue ***(McDicken, 1991).***

Acoustic Shadowing and Artifacts:-

In biologic tissues the speed of the sound is lowest in gas, faster in fluid, and fastest in bone, where the molecules are more closely packed. The sound pulses transmitted into the body can be reflected, scattered, refracted or absorbed. Absorption or attenuation is the loss of acoustic energy by conversion to heat energy, more prevalent in bone than soft tissue, and more prevalent in soft tissue than in fluid. It is a key cause of acoustic shadowing. Where there is a distinct loss of the echoes behind an imaged structure. Acoustic shadowing is so common in ultrasound images that it is sometimes called an artifact. It is the result of the energy (of transmitted sound) that is being decreased by reflection and/or absorption. The shadowing behind gas is due to strong reflections at gas/tissue interfaces. The reflected pulse interacts with interfaces in front of the gas causing secondary reflections, which leads to low level echoes, causing ‘dirty’ images. However, the shadowing that occurs behind stones, calcifications and bones is reduced by sound absorption, resulting in only minimal secondary reflection, and therefore ‘clean’ images ***(Middleton et al., 2004).***

While performing ultrasound images of a liver, one may see multiple, vertical, long, narrow bands or lines extending down from the posterior surface of the right hemidiaphragm. These are ring-down artifacts. These findings have been noted to be most prevalent in patients with emphysema, idiopathic interstitial pneumonia, bronchopneumonia and interstitial edema. It is speculated that the ring-down arises from thickened intralobular or interlobular septa filled with fluid touching the visceral pleural surface ***(Lim et al., 1999).***

The comet tail artifact is a reverberation artifact; reverberation artifacts are strong reflections, in multiples, from the same surface. These artifacts look similar to ringdown. The comet tail artifact is an antishadow, a trail of dense continuous echoes simulating a comet tail. They are usually associated with foreign bodies, especially metallic objects such as surgical clips, and cholesterol foci ***(Avruch and Cooperberg, 1985).***

The more the acoustic impedance of the object differs from the surroundings, the greater the number of reverberant echoes. The smaller the object is, the closer is the spacing between these echoes. If echo bands are strong and close together, they merge to produce the comet tail pattern. The reverberation is strongest when the object is perpendicular to the ultrasound beam. In comparison to the ringdown artifact, the comet tail artifact tapers fast and is short. Posterior enhancement occurs when fluid-containing structures attenuate the sound less than solid structures, the strength of the sound pulse increasing after passing through fluid compared to passing through a solid structure. This increase through transmission distinguishes cysts and fluid collections from solid masses ***(Ziskin et al., 1982).***

Echogenicity Ultrasound images are displayed on a gray scale. The strongest echo appears white while it is black when no sound wave is reflected from the organs. Depending on the reflected wave amplitude, the following terms are used to define echogenicity. When no sound wave is reflected and the image appears black it is *anechoic* as in pleural effusion. It is *isoechoic* when the echoes are of comparable amplitude with the surrounding tissue as with kidneys or spleen. It is *hyperechoic* when echoes are stronger than the surrounding tissue as in diaphragm, and *hypoechoic* when it is weaker than that from the surrounding tissue ***(Ziskin et al., 1982).***

Diagnostic Ultrasound Equipments:-

Regarding diagnostic ultrasound equipment, ceramic crystals in the transducer deform and vibrate when electronically stimulated to produce the sound pulses. Echoes that return to the transducer distort these crystal elements and produce an electric pulse, which is processed into an image. High-amplitude echoes create greater crystal deformation and produce a larger electronic voltage. Resolution of an image is very important for diagnosing pathology. Resolution is determined by the frequency and duration of the transmitted sound pulse. Axial resolution refers to the ability to resolve objects within the imaging plane at different depths along the direction of the pulse, best with higher frequency probes with their shorter pulses. Lateral resolution is the ability to resolve objects in the imaging plane that are located side by side ***(Bushberg and Seibert, 2002).***

The pulsed ultrasound energy is controlled by the system's electronics and emitted from the probe or transducer. The probe can have a single element or be composed of an array of many small elements that can be individually addressed and controlled. The latter is referred to as a phased array transducer. The elements are used both to transmit the ultrasound as well as to detect the energy directed back towards them ***(Bushberg and Seibert, 2002).***

It should be noted that the individual elements cannot simultaneously transmit and receive so following the emission of a pulse, an element in the probe or transducer starts listening for the echo. After a sufficient amount of time has passed, corresponding to a certain desired depth for acquiring information for an image, another pulse can be emitted. The size of the transmitted beam is related to both the size and number of elements that are used. By controlling the timing of transmission from individual (or groups of) elements, the ultrasound beam's direction and focusing can be controlled to obtain the image of the organ being studied. Most transducers today are multielement probes called arrays. They contain groups of small crystal elements in a sequential linear fashion.

By changing the timing and sequence of activation of the different arrayed elements, the pulse can be directed to different places and focused at specific depths depending on the organ being interrogated. In the phased array transducer each element in the array helps in the formation of each pulse. A sector image is created by this probe, which is small and can fit between ribs ***(Bushberg and Seibert, 2002).***

Curved-array transducers have a convex shape for a wider field of view. A 3.5-MHz curvilinear probe provides visualization of deeper structures, and the sector scan field allows a wider field of view through a small acoustic window. These transducers are required to image a thicker thoracic wall. The chest wall, pleura, and lungs can be evaluated using this probe. The posterior chest is best imaged with the patient sitting upright, the anterior and lateral aspects of the chest in the lateral decubitus position. The suprasternal approach is the best way to view the upper anterior and middle mediastinum; the aorta and superior vena cava can be seen ***(Koh et al., 2002).***

In abdominal and pelvic imaging curved-array transducers are used to image the general abdomen including the pelvis for obstetrical cases. Curved array transducers with a short radius can be used as intraluminal or endoluminal probes. These transducers are small and can be positioned close to the organ of interest, use higher frequencies, and thus obtain higher resolution for very detailed images. Not having to transmit sound through the abdominal wall minimizes the major degradation by adipose tissue. With linear-array transducers (a limited group of adjacent elements produce a pulse), which is perpendicular to the transducer face ***(Middleton et al., 2004).***

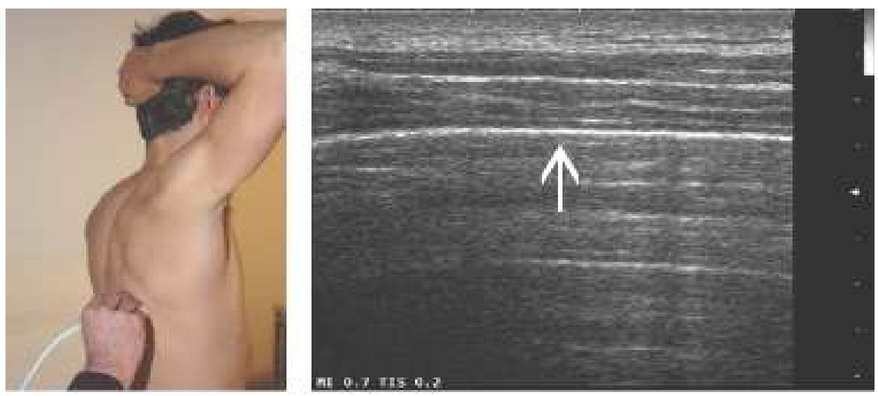
The image is rectangular. Line array transducers of high frequency are well-suited for patients with thin thoracic walls. The major benefit of this large transducer is high resolution in the near field and a large superficial field of view ***(Meuwly and Gudinchet, 2004).***

The newer harmonic imaging uses higher integer multiples of the fundamental transmitted frequency. These sound waves progressively increase in intensity before they attenuate. A filter allows only the high-frequency harmonic signal to be processed into an image ***(Middleton et al., 2004).***

Harmonic imaging sometimes provides a smoother-appearing image. Three dimensional (3D) sonography data are acquired as a stack of parallel cross sections with a 2D scanner or as a volume with a mechanical or electronic-array probe. 3D images are used selectively to better appreciate the shape of a mass or organ and its relationship to surrounding structures ***(Middleton et al., 2004).***

Examination Technique:-

Usually the dorsal and lateral images are obtained with the patient sitting **Fig. (1)**, whereas the supine position is used for visualizing the ventral side. Raising the arms and crossing them behind the head causes intercostal spaces to be extended and facilitates access. The examiner is able to visualize the region behind the shoulder blade, if the patient puts his/her hand on the contralateral shoulder. The transducer is moved along the intercostals space from dorsal to ventral in longitudinal and transversal positions. Turning the probe in different positions provides the examiner with a three-dimensional image. During every stage of examination, the user should determine the breath-related moving of the pleura, the so-called sliding sign ***(Middleton et al., 2004).***



a b

Fig. (2): a: Linear probe placed intercostally in an oblique view. The right arm is elevated behind the head. b: corresponding sonographic view "sliding line of the visceral pleura ***(Middleton et al., 2004).***

From the abdomen, in subcostal section by the transhepatic route on the right side **Fig. (3)** and to a lesser extent through the spleen on the left side, the diaphragm is examined. The axilla should be examined in the supine position with the arm abducted over the head. The supraclavidular acces allows the investigator to view the region of the brachial plexus, the subvlavian vessels and the lung tip. From suprasternal, the anterior upper mediastinum can be viewed. Bed ridden and intensive care patients are examined by turning them to the oblique position in the bed ***(Middleton et al., 2004).***

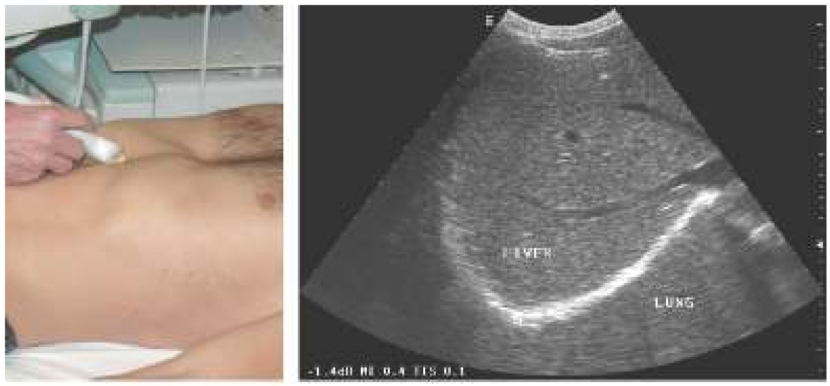


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Fig. (4): a: Convex probe placed subcostally from the right. b: Corresponding sonographic image, Lung is indicated as a mirror artifact above the diaphragm ***(Middleton et al., 2004).***

How Can Diagnostic Ultrasound Help the Pulmonologists?

Diagnostic ultrasonography is a very valuable tool for imaging the chest because it causes no clinically significant biological effects, is a real-time examination and has multi planar imaging capability. In real time one can focus the study on a painful or palpable area. This modality of ultrasonography can be portable, very significant for the ICU and emergency room. Transthoracic ultrasound can be used to evaluate peripheral parenchymal, pleural and chest wall diseases. The maximum visualization of the lung and pleural space is done by scanning along the intercostal spaces during quiet respiration for normal lung movement; and in suspended respiration when a lesion can