The (CDH) Scoring System

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Three main clinical presentations of (CDH) can be observed:-

Group-1:
A group of patients with severe cyanosis from the moment of delivery which is totally unresponsive to treatment because of insufficient lung for effective gas exchange and death tends to occur rapidly.
Congenital Diaphragmatic Hernia (CDH)

- **Group-2**: Patients present with respiratory distress and cyanosis within the first hour or two of extra-uterine life, but with treatment can attain adequate PaO2. These patients have enough lung to allow gas exchange, but have a very liable pulmonary circulation as a result of pulmonary hypertension, (delayed surgical group).
Congenital Diaphragmatic Hernia (CDH)

- **Group-3**;

A group that present with more minor respiratory distress on feeding or at rest or may be detected as having dextro-cardia on routine x-ray examination.

This group usually do not cause the usual dilemma in (CDH) management.
Congenital Diaphragmatic Hernia (CDH)

• **In sonography**, any abnormality resulting in either a solid mass or cyst within the fetal thorax can be confused with Cystic Adenomatoid Malformation (with all of its types), Pulmonary Sequestration, Bronchogenic Cyst, and Terratomas, may all simulate CDH,

• Likewise, CDH, may simulate these lesions, therefore it should be remembered that these lesions may often coexist making diagnosis quiet difficult.
Congenital Diaphragmatic Hernia (CDH)

- However, Antenatal (USS) as well as pre-op. x-ray scoring system can be as reasonably accurate as (ABG) values in risk assessment of babies with (CDH) anti-natally diagnosed,
- All together may give the surgeon another way of identifying the highest risk patients and determine the effective method of management and the reasonable time of surgical intervention.
Congenital Diaphragmatic Hernia (CDH)

- The Non-responders are those with post-ductal PaO2 never rises above 60 torr. These at highest risk children are symptomatic either in the delivery room or within 6 hours after birth. With severe persistent post-ductal hypoxemia, have fixed right-to-left shunt, un-responsive to efforts aimed at improving pulmonary blood flow.
Congenital Diaphragmatic Hernia (CDH)

- The Non-responders pulmonary vascular bed may be inadequate to support a normal cardiac output, and they shunt a large percentage of their right ventricular output into the descending aorta. This leads to progressive arterial desaturation.

- (Greater measures here are unsuccessful, only surgery could be tried).
Congenital Diaphragmatic Hernia (CDH)

- The **Responders** are those with post-ductal PaO2 of 91-to-500 torr.
- Those are who enter the “honey moon period.”
- They have an abnormally labile and reactive pulmonary vasculature.
- Their lungs however, hypo-plastic, demonstrate the ability to accept a full or even excessive cardiac output.
Congenital Diaphragmatic Hernia (CDH)

- These Responders have normal pulmonary vascular resistance with minimal hypertension and can maintain normal oxygenation,
- However, many events including suctioning of the endo-tracheal tube, loud noises, painfull procedures and atelectasis can trigger intense pulmonary vasoconstriction with resultant right-to-left shunting.
Congenital Diaphragmatic Hernia (CDH)

- If appropriate measures are not instituted, (paralysis, sedation) these Responders may also die.
- (It is this group that seems to respond to other efforts of prolonged management, waiting and delayed surgery).
Congenital Diaphragmatic Hernia (CDH)

- **Contemplations :-**
  1/ Can the Non-responders be identified by PaCO2 ventilation indexes, arterial PaO2, and A-a gradient without aggressive manipulation of a critically ill newborn?
  2/ Has our approach statistically changed the survival data in CDH when compared to historical controls managed more conventionally?
  3/ Despite advances in neonatal resuscitation and intensive care in the past two decades the mortality rate remains frustratingly high?
Congenital Diaphragmatic Hernia (CDH)

- There is no doubt that surgical repair of CDH is often the easiest part of the overall management of these newborns.
- **The cause of the high mortality is their associated pulmonary hypoplasia with its two primary components:**
  1. Reduced surface area for gas exchange [this is maximum in gr-1. Severe hypoplasia]
  2. Increased muscularity of the pulmonary arteries and their branches. (v. sensitive to various stimuli).
• The resulting arterial constriction results in pulmonary hypertension (PHT),

• And the subsequent shunting of de-saturated blood to the systemic arterial circulation through the ductus arteriosus (PDA), the foramen ovale (FO), and intrapulmonary (A-V) communications.

• Thus the neonates circulatory anatomy reverts to that of the fetus (Persistent Fetus Circulation-PFC).
Congenital Diaphragmatic Hernia (CDH)

- Initially many neonates may achieve normal or even supra-normal (PaO2) values, demonstrating adequate lung surface area for gas exchange,
- This situation is usually short lived, as it is followed in gr-2 neonates, by hypoxia and acidosis secondary to PFC.
- Unless active measures are taken the neonate will die.
- Some gr-3 neonates will also develop PFC but this is not usually severe.
Congenital Diaphragmatic Hernia (CDH)

- **Polyhydramnios**, was not only common, but was also a predictor of extremely poor clinical outcome as high as 5x than those fetuses without polyhydramnios,

- **Polyhydramnios** is a good prenatal marker for the presence of CDH, and a good predictor for severe cases with poor prognosis.
Congenital Diaphragmatic Hernia (CDH)

- The (CDH) Scoring System:

1. The Presence of Polyhydramnios in association with a fetus with CDH is a predictor of poor outcome with a mortality rate of about 90%.

However, this is a nonspecific sign as there are many other causes of increased liquor volume,

- Present ----------- 0
- Not present ------ 2
Congenital Diaphragmatic Hernia (CDH)

- **The (CDH) Scoring System:**

  2 - *APGAR Score*; (ante-natally diagnosed as having CDH),

  - 1st, 5th, and 8th minute Apgar scores are recorded and according to the total score, 0, 1, or 2 points are scored, (the lower Apgar, the lower the score).

  = A------ 0
  = B------ 1
  = C------ 2
Congenital Diaphragmatic Hernia (CDH)

• **The (CDH) Scoring System**: 

3 - Gestational Age;

- < 30 weeks of gestation-------- 0 pts
- > 30- <36 weeks of gestation-- 1 pt
- > 36 weeks of gestation-------- 2 pts

Preterm newborns have the least chance of survival because of expected lung immaturity even without the effect of CDH.
Congenital Diaphragmatic Hernia (CDH)

• The (CDH) Scoring System :-

  4 - Birth Weight ;

  < 1000 g --------------------- 0 points
  > 1000g but <2000 g ----- 1 point
  > 2000g ------------------------ 2 points

(VLBW) babies have 0 score. The (LBW) has lesser chance of survival than the > 2kg neonates with CDH.
Congenital Diaphragmatic Hernia (CDH)

The (CDH) Scoring System:

5 - Age at Onset of Symptoms;

- Prognosis in individual cases may be predicted by the age at onset of symptoms,
- Presentation under 2 hours of age with respiratory symptoms implies significant pulmonary hypo-plasia and high mortality,

= < 2hrs of age-------- 0
= 2 to 6 hrs of age------ 1
= > 6 hrs of age-------- 2
Congenital Diaphragmatic Hernia (CDH)

- The (CDH) Scoring System:
  - The relationship between blood gas values (BG), mechanical ventilation and the degree of lung development.
  - Immediately after first Apgar, after transfer to NICU, before and after X-ray taking,
  - Various attempts have been made to predict outcome based on (pH, PaO2, PaCO2, A-aDO2).
Congenital Diaphragmatic Hernia (CDH)

- The (CDH) Scoring System:

  6 - ABG Values with high mortality and unlikely survival;

  * pH .............. < 7.0
  * PaCO2 ........... > 60 mmHg
  * PaO2 ............ < 50 mmHg
  * Aa-DO2 ........... > 500 torr on 100% O2

  = ------------------0 (points)
  = ------------------1 (pH > 7.0 and improves)
  = ------------------2 (pH well > 7.0 and maintain)
Congenital Diaphragmatic Hernia (CDH)

• A pre-reduction alveolar to arterial oxygen pressure difference (A-aD\text{O}_2) of over 500 torr obtained during 100% oxygen breathing is associated with high mortality rate.

• Therefore poor prognosis is expected when severe respiratory acidosis does not improve after positive pressure mechanical ventilation and is associated with a high degree of A-V shunting.
Congenital Diaphragmatic Hernia (CDH)

- The (CDH) Scoring System:
- 7- Vasodilator Therapy:
  - There are no selective pulmonary vasodilators,
  - These agents may be harmful,
  - By increasing vascular capacity their use results in a large I.V. volume requirement,
  - This increase in total body and interstitial lung water, results in higher inspiratory pressures being required for adequate ventilation, and this may impede pulmonary blood flow.
Congenital Diaphragmatic Hernia (CDH)

- The (CDH) Scoring System :-
- 7- Vasodilator therapy ;
  - The use of vasodilators may improve oxygenation slightly but not the overall survival. (Response to Tolazoline as a predictor of outcome) ;
  =-----------------0 (NO Response)
  =-----------------2 (+ve Response)
Congenital Diaphragmatic Hernia (CDH)

- Preoperative X-Ray Scoring :-
  - Favorable and unfavorable x-rays and their cumulative score,
  - To substantiate the diagnosis,
  - Rule out other possibilities like lung (CCAM),
  - Monitor n.g. tube position, (if it is already put.),
  - Monitor e.t. tube position, (if it is already put.),
  - And detect a pneumothorax (if any).
Congenital Diaphragmatic Hernia (CDH)

Different types of Congenital Diaphragmatic Hernia
Congenital Diaphragmatic Hernia (CDH)

- **Preoperative X-Ray Scoring:**
  - As survival is principally influenced by pulmonary function, those babies with the most visible aerated lung and those with the least pulmonary compromise from compression of lung by dilated loops, stomach, or pneumothorax would be expected to have the best prognosis.
Congenital Diaphragmatic Hernia (CDH)

- Preoperative X-Ray Scoring:

- Each chest x-ray finding is graded on a sliding scale from zero to two with the higher grades assigned to the favorable findings,

- The sole exception is assigning a grade of (one) out of (three) to a right sided CDH, containing liver.
Congenital Diaphragmatic Hernia (CDH)

- **Preoperative X-Ray Scoring**: -
  - In order to plan postnatal treatment, a reliable and easily identifiable antenatal marker predicting the severity of pulmonary hypoplasia was required, *(the thoracic site of the stomach diagnosed ante-natally and confirmed by postnatal radiographs)*,
  - This finding is then correlated with mortality due to PFC defined as right-to-left shunting confirmed by pre- and post-ductal PaO2 measurements.
Congenital Diaphragmatic Hernia (CDH)

• **Preoperative X-Ray Scoring :-**
  - The presence of thoracic stomach is simply a marker of intestinal herniation early in fetal life, while abdominal stomach indicates later herniation,
  - Early herniation may result in more severe lung compression and an earlier arrest in lung maturation,
  - Therefore, it is possible that, most if not all neonatal diagnosis of left (CDH) will be made when the stomach is seen to lie in the chest.
Congenital Diaphragmatic Hernia (CDH)

CDH:
- Stomach in the chest
- Demonstrated by nasogastric tube position (white arrow).
Congenital Diaphragmatic Hernia (CDH)

• Preoperative X-Ray Scoring :

1- Side of Diaphragmatic Hernia ;

= Right with liver-------------------------- 1
= Right------------------------------------ 2
= Left------------------------------------- 3
Congenital Diaphragmatic Hernia (CDH)

- **Preoperative X-Ray Scoring**:
  - 2 - Stomach Location;
    - Chest: 1
    - Abdominal: 2
Congenital Diaphragmatic Hernia (CDH)

- **Preoperative X-Ray Scoring** :-

  **3 – Pneumothorax** ; (Ipsilateral or contralateral)

  - Present 0
  - Absent 2
Congenital Diaphragmatic Hernia (CDH)

- Preoperative X-Ray Scoring:

  4 - Degree of Mediastinal Shift:

  - Marked: 0
  - Mild: 1
  - None: 2
Congenital Diaphragmatic Hernia (CDH)

- **Preoperative X-Ray Scoring :-**

  5 - Amount of Visceral Distension ;
  
  - Distended -------------- 0
  - No Distension -------- 1
  - No Air --------------- 2
Congenital Diaphragmatic Hernia (CDH)

- **Preoperative X-Ray Scoring**: 
  
  6 - Volume of Areated Ipsilateral Lung ;

  \[
  = 00\% \quad \text{---------} \quad 0 \\
  < 25\% \quad \text{---------} \quad 1 \\
  > 25\% \quad \text{---------} \quad 2
  \]
Congenital Diaphragmatic Hernia (CDH)

- Preoperative X-Ray Scoring:

  7- Volume of Areated Contralateral Lung:

  - 50 to 25% 0
  - 75 to 50% 1
  - Normal to 75% 2
Congenital Diaphragmatic Hernia (CDH)

• Using (5) Five of the (7) Seven Roentgen findings, significant correlation with survival can be obtained as a total x-ray score:

- Non-Survivors Score ≤ 6 (< 6),
- Survivors Score ≥ 6 (> 6).
Congenital Diaphragmatic Hernia (CDH)

- **Observed Primary Ventilation Results:**
  
  (Ventilatory predictors of pulmonary hypoplasia and the effect of surgical repair on respiratory mechanics). *(Survivors / Non-survivors).*

1. Patients with $\text{PaCO}_2 < 40\text{mmHg}$ or $(>40\text{mmHg})$,  
2. Whose lungs are easy to hyperventilate or *(there is resistance).*  
3. Mean airway pressure *(MAP)* $< 20 \text{cm H2O}$, or $(> 30 \text{cm H2O})$,  
4. Respiratory rate *(RR)* $< 60/\text{min}$, or $(> 60/\text{min})$.  

Congenital Diaphragmatic Hernia (CDH)

- **Arterial CO2 :-**
  - Accurately reflects the degree of lung development in (CDH), and separates those patients with severe pulmonary hypo-plasia where the outcome is invariably fatal, from those with a well developed contra-lateral lung where there is excellent potential for survival.
Congenital Diaphragmatic Hernia (CDH)

- Patients with elevated PaCO2 levels which can not be reduced by hyperventilation, have bilateral pulmonary hypo-plasia which is associated with severe pre-ductal shunting and death.
- Other group of patients that respond well to hyperventilation with reduction in PaCO2, has ductal shunting, but invariably survivors.
Congenital Diaphragmatic Hernia (CDH)

- **Echocardiography and (PH) :-**
  - M-mode echocardiography is useful in early identification and monitoring of newborns with (PH) and clinically significant (PFC),
  - Pulmonary vascular resistance can be evaluated non-invasively as newborns make the transition from fetal to mature lung physiology and normal pulmonary vascular resistance.
Congenital Diaphragmatic Hernia (CDH)

- **Echocardiography and (PH) :-**
  - The clinical correlation seen suggests a role of echocardiographic monitoring in:
    - Evaluating persistence of (PH),
    - Following therapeutic response, and
    - Documenting final stabilization of the pulmonary vascular bed before medical support withdrawal.
Congenital Diaphragmatic Hernia (CDH)

- Echocardiography and (PH):-

  - The (RVSTI), Right Ventricular Systolic Time Interval, as measured from an echo and electro-cardio-graphic tracing (normal newborn value 0.34 +/- 0.06 by 6 hrs or more of age), has shown good correlation with diastolic pulmonary artery pressure and pulmonary vascular resistance.
Congenital Diaphragmatic Hernia (CDH)

• **Other predictors of survival:**
  • 5 clinical parameters derived from neonates best preoperative ventilatory and BG data in the first 24 hrs of life;
    • 1- PaO2
    • 2- PaO2 : FIO2
    • 3- A-a DO2
    • 4- OI (Oxygenation Index) = MAP x FIO2 / PaO2
    • 5- MVI (Modified Vent. Index) = PIPxRRxCO2 / 1,000
Congenital Diaphragmatic Hernia (CDH)

Other Predictors of Survival :

- Criteria for inclusion ;
  - 1 - CDH diagnosed within 6 hours of delivery,
  - 2 - Ventilation before repair,
  - 3 - No associated lethal cong. abnormalities.
Congenital Diaphragmatic Hernia (CDH)

- **Parameters Analysed** :-
  - OI of > 0.08 predicted a 94% chance of survival with ;
    - a sensitivity of 96%
    - a specificity of 95%
  - MVI of > 40 predicted a 91% chance of survival with ;
    - a sensitivity of 94%
    - a specificity of 86%
Congenital Diaphragmatic Hernia (CDH)

• Three Groups Identified:

  (OI and MVI values derived from their best preoperative ABG):

  ❖ 1- Those with an OI of (> 0.006) or an MVI of (> 40) can be expected to survive with conventional therapy of 98% and 96% accuracy respectively. They are likely to have minimal pulmonary hypoplasia.
Congenital Diaphragmatic Hernia (CDH)

- Present with minor respiratory distress on feeding or at rest or may be detected as having dextro-cardia on routine examination as an accidental finding,
- All are supposed to survive.
2- Those with OI < 0.175 and MVI < 80 ;

All can be expected to die with conventional therapy. These newborns are likely to have a degree of lung hypo-plasia in-compatible with life, and are unlikely to benefit from therapies designed to combat pulmonary hypertension and minimize baro-trauma.
Congenital Diaphragmatic Hernia (CDH)

- Present with severe cyanosis from the moment of delivery,
- They are totally unresponsive to treatment,
- In this group there is insufficient lung for gas exchange,
- Death always occurs rapidly.
Congenital Diaphragmatic Hernia (CDH)

3- Between those two groups (extremes), is a small group of neonates (OI and MVI are between those extremes). They have more uncertain outcome with conventional therapy. This group may well have significant (PH) and moderate pulmonary hypo-plasia and may as well survive using ECMO specifically designed to treat (PH) and minimize lung injury.
Congenital Diaphragmatic Hernia (CDH)

- Present with respiratory distress and cyanosis within the first hour or two of life, but with treatment can attain adequate PaCO2.
- These newborns have enough lung to allow gas exchange but have a very labile pulmonary circulation as a result of pulmonary hypo-plasia.
Congenital Diaphragmatic Hernia (CDH)

• More recent efforts have centered upon assessing prognosis by virtue of the (amount of identifiable pulmonary parenchyma on the contra-lateral lung),

• To achieve that two techniques (measuring right lung area and comparing the subjective assessment of the right lung area to the right haemithorax area) appear easy to perform and reasonably accurate.
Congenital Diaphragmatic Hernia (CDH)

• **First Technique :-**
  - At the level of the atria on a transverse axial scan, of the fetal thorax, right lung area is measured,
  - **Measured Rt lung area : Head circumference ratio**, 
  - All fetuses with ratio < 0.6 died. Survival was 100% when the ratio was > 1.35,
  - Two-dimensional perpendicular linear measurement in millimeters was used to calculate the area.
Second technique:

- Lung area was determined on a transverse axial plane at the level obtained to visualize the four cardiac chambers and the hemi-thorax area is obtained by drawing a posterior line tangential to the border of the vertebral body on the same side of quantified lung area,

- Fetuses with **Lung Area to Hemi-thorax > 50%** had a **86%** survival, while ratio **< 50%** had **25%** survival.
Congenital Diaphragmatic Hernia (CDH)

- **Bohn’s Quadrant Scoring Criteria; Mortality%**;
  
  (VI = Ventilation Index = Mean Airway Pressure x Ventilatory Rate)

- **A)**- PaCO2 > 40 + VI < 1,000 70%
- **B)**- PaCO2 < 40 + VI < 1,000 14%
- **C)**- PaCO2 > 40 + VI > 1,000 100%
- **D)**- PaCO2 < 40 + VI > 1,000 57%