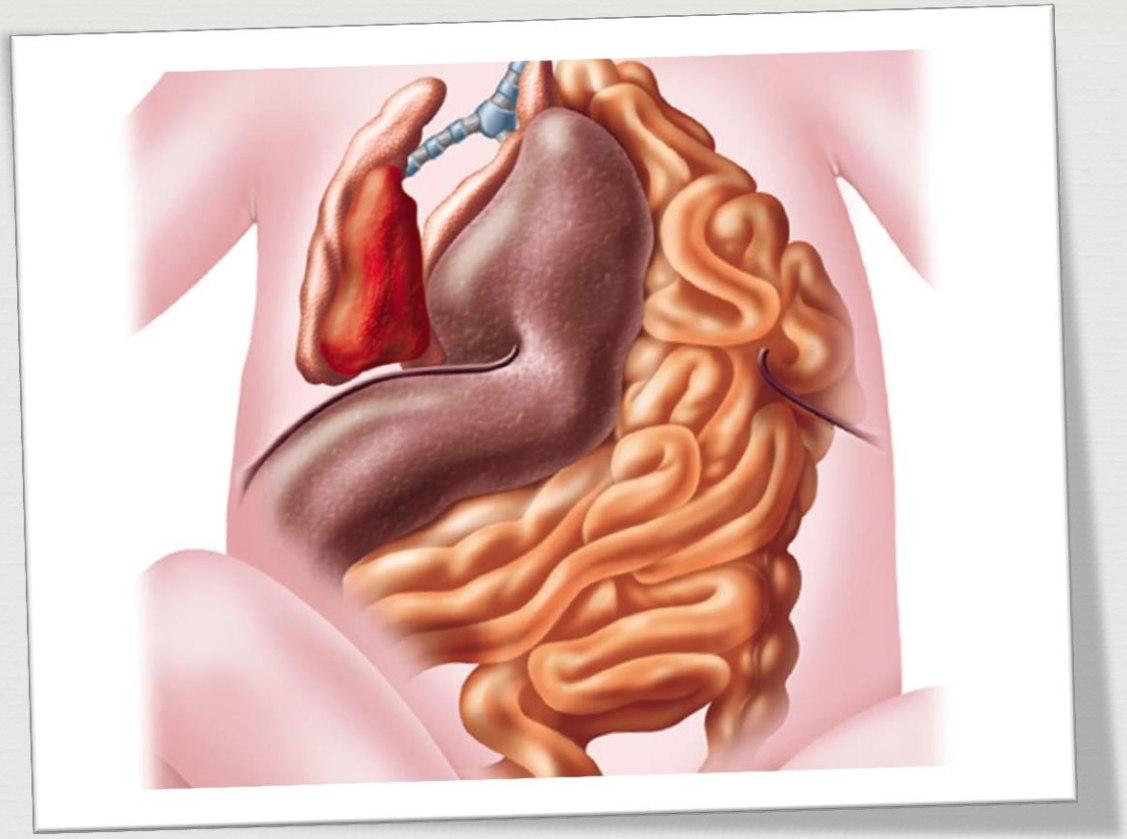
A RESEARCH JOURNEY- ENHANCING THE MANAGEMENT OF CONGENITAL DIAPHRAGMATIC HERNIA

Alana J Coleman, BSN, RN, CCRN

Department of Neonatology
Children's Hospital of Pittsburgh of UPMC
Pittsburgh, Pa, United States



WHAT IS A CONGENITAL DIAPHRAGMATIC HERNIA?

Congenital Diaphragmatic Hernia (CDH) is a congenital defect of the diaphragm that results in an abnormal opening in the diaphragm that allows bowel loops and other abdominal organs to herniate into the chest cavity.

Congenital Diaphragmatic Hernia: Updates and Outcomes. Jennifer R. Benjamin, MD, Matthew J. Bizzarro, MD and C. Michael Cotten, MD, MHS. NeoReviews Vol. 12 No. 8 August 1, 2011. pp. e439 -e452

CDH: PROGNOSIS

- PULMONARY HYPOPLASIA
  - Fewer airways and alveoli
    - Decreased surface area for gas exchange: poor oxygenation
- PULMONARY HYPERTENSION
  - Vascular Hypermuscularity
    - Fixed vascular resistance leads to pulmonary hypertension
- ASSOCIATED ANOMALIES (40-50%)
  - Heart disease (20%) remains significant risk factor for death in patients with CDH
  - Chromosomal anomaly (5-15%)
  - Other: renal, neural tube defects, gut malrotation

Congenital Diaphragmatic Hernia: Updates and Outcomes. Jennifer R. Benjamin, MD, Matthew J. Bizzarro, MD and C. Michael Cotten, MD, MHS. NeoReviews Vol. 12 No. 8 August 1, 2011. pp. e439 -e452

CDH: THE IMPACT

- Epidemiology
  - Estimated 1:2,400 live births
  - No known association with gender
- Mortality
  - National Survival rate: 74%
  - Population-based studies: 50-92%
- Cost
  - Estimated cost per new case: $757,575
  - Overall estimated yearly cost: $800,000,000

Congenital diaphragmatic hernia- the past 25 (or so) years. Lally, K. J Ped Surg. 2016; 51, 695-698

CASE PRESENTATION



February 2005

(Left CDH)

BIRTH HISTORY

- Vaginal delivery, 39 weeks, 3.05 kg
- APGAR scores: 5,8
- Spontaneous resp effort and crying at birth
  - Intubated within 3 minutes
  - R 50 32/6 100%
- Initial ABG: 6.97/102/46/22-8
  - HZ 10 AMP 26 MAP 16 NO 20
  - Vecuronium/NAHCO3
  - CXR: R pneumo
    - CT placed

NICU

- 7.19/35/69/26-4 SPO2 100/84
  - HZ 8 AMP 30 MAP 18 100% 20 NO
- Medications:
  - Vecuronium/Fentanyl/NAHCO3/Dopamine gtts
- Dad to bedside
  - 50% chance of survival

COURSE

- ECHO: Systemic RV pressure, R-L PDA, PFO vs ASD, Decreased LV function
  - Dobutamine and dopamine
- 5 days: HZ 7 AMP 32 MAP 18 FIO2 90 NO 20
- Repair: Subcostal, PATCH (gortex)
  - Bowel, liver, spleen, stomach
  - L CT

OUTCOME

- 6 hours post-repair: hypotensive, pulm hemorrhage
- Arrested 30 minutes
- Cannulated emergently onto ECMO
- EEG: Burst suppression, increased electrographic sz
- Repeat EEG :Anoxic brain injury
- Support withdrawn

Personal stories are often the catalyst for immediate action in the world of healthcare

Larin, L.R. (2013). Inspired to Change: Improving patient care one story at a time. Chicago, IL: Health Administration Press.

KAYS, D.W.

- OUTCOMES
  - ERA 1 (1983-1998): aggressive ventilation, no ECMO
    - Survival 15%, 17% treated patients
  - ERA 2(1998-1992): aggressive ventilation, ECMO
    - Survival 44% overall, 50% treated
  - ERA 3 (1992-present): gentle ventilation, ECMO
    - Survival 78% overall, 89% treated patients

Detrimental effects of standard medical therapy in congenital diaphragmatic hernia. Kays, D.W. et al., Ann Surg. 1999; 230, 340-348

QI STUDY



Management of Congenital Diaphragmatic Hernias

OBJECTIVE

- Do we need to change CDH protocol?
- Collect and evaluate data from the last 20 CDH patients
- Compare management strategies from hospitals who implement permissive hypercapnea/spontaneous respiration/delayed repaired approach

BACKGROUND

- 20 PATIENTS (2-12-03 TO 4-5-05)
  - 17 LEFT, 2 Right and 1 bilateral
  - 5 (25%) pre-op pneumothoraxes
  - 9 (45%) required ECMO
    - 4 arrested onto ECMO
  - 50 % overall survival

20 Patients reviewed

	SURVIVORS	NONSURVIVORS
Gest Age	33.5	37
Weight	2.83	2.83
Prenatal DX	78%	80%
Pre op pneumo	10 %	40%
ECMO	20%	70%
HFOV	50%	100%
Nitric Oxide	50 %	100%
Survanta	10%	20%
Steroids	60%	70%

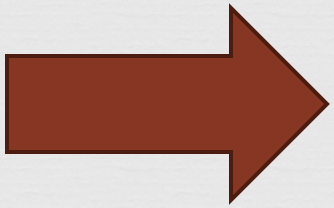
HYPERVENTILATION - More harm than good

- 1981- Drummond and colleagues demonstrated with either hyperventilation or alkalosis, patient oxygenation increased due to relaxation of pulmonary vasculature
- 1995- JT Wung reports use of low pressure ventilation in infants with pulmonary hypertension and the CDH population
- 1999- Series of reports demonstrated gentle ventilation, permissive hypercapnia strategy

Congenital diaphragmatic hernia- the past 25 (or so) years. Lally, K. J Ped Surg. 2016; 51, 695-698

CDH: EVOLUTION OF MANAGEMENT

- Hyperventilation
- Alkalization
- Sedation/Paralysis
- Early Operation
- Emergent ECMO



- 'Gentle Ventilation'
- Permissive Hypercapnia
- Spontaneous respiration
- Delayed Surgery
- Elective ECMO

SURGERY- does timing matter?

- 1980's –CDH was the most urgent acute surgical emergency in the newborn infant.

- Reports for delayed surgical intervention surfaced from Sakai and Langer, as well as Hazebroek from the Netherlands, emphasizing the utilization of perioperative stabilization.

Congenital diaphragmatic hernia- the past 25 (or so) years. Lally, K. J Ped Surg. 2016; 51, 695-698

ECMO

- Used as *preoperative rescue therapy* for those with severe hypoplasia or PPHN
- Success dependent on reversibility of condition
- Allows delayed surgical approach for severe cases

Congenital diaphragmatic hernia-stabilization and repair on extracorpeal membrane oxygentaion. K.P.Lally, M.S. Paranka, J Roden, et al. Ann Surg 1992; 216, 569-573

RESULT

- Unanimous decision amongst pediatric surgery and neonatology to change CDH protocol to entail permissive hypercapnea/spontaneous respiration/delayed repair approach

- Multidisciplnary team emerged to develop new protocol

TEAM

- NURSING
- PEDIATRIC SURGEON
- NEONATOLOGIST
- RESPIRATORY THERAPIST
- PERFUSION

EDUCATION

- *Everyone* who has an encounter with this patient will be touched.

- And everyone who touches the patient should have advanced knowledge of what is happening.

https://ecmoadvantage.com/there-is-no-i-in-team/

Aim of therapy is to achieve the Goal Cardiorespiratory Parameters while maintaining patient support in the Comfort Zone

If infant **exceeds comfort zone** therapies to maintain the goal parameters than Neonatology and Pediatric Surgery attendings will discuss time limits for current therapies and endpoints leading to ECMO support

Goal Parameters

- Preductal PaO2 goal 60–80
- Preductal SaO2 goal >85%
- SvO2 saturations >60%
- PCO2 goal <65–75
- pH goal 7.20–7.25
- Lactate goal <3 mmol/L
- UOP goal <1.5 ml/kg/hr

Comfort Zone

- CMV PIP <25
- HFOV MAP <17
- Dopamine infusion 10-15 mcg/kg/min
- Hydrocortisone (50mg/m2)
- Inhaled Nitric Oxide (20ppm)

Congenital Diaphragmatic Hernia *UPMC Clinical Management Guidelines 2007*

IRB STUDY

- The study objectives for **"Therapeutic Interventions, Management and Outcomes of Congenital Diaphragmatic Hernia (CDH) Patients: An Increase in Survival"** were met. We did evaluate 47 neonates admitted to our institution with the diagnosis of CDH from **January 2007-December 2010 (overall survival 64%)**. We looked at the years each infant was born and assigned the SNAP-II score to assess severity of illness. By calculating the Snap-II scores we were able to predict which of these specific patients required ECMO therapy and which infants were more likely to die. Our findings were significant and were presented at the 27th Annual CNMC Symposium on ECMO and Advanced Therapies for Respiratory Failure conference in February 2011 and were finally published in the Journal of Pediatric Surgery in November of 2013.

Distribution of Clinical Characteristics: ECMO Vs. No ECMO

Variables	ECMO n = 24	No ECMO n = 23	P Value
Gestational Age (weeks)	38.50 [37.0 – 39.0]	38.0 [36.0 – 39.0]	0.25
Weight (kg)	3.04 [2.62 – 3.31]	2.90 [2.45 – 3.22]	0.36
Male Gender	13 (54.2%)	16 (69.6%)	0.22
Median APGAR (5 min)	7.00 [4.0 – 8.0]	8.0 [6.0 – 8.0]	0.33
Right-Sided Defect	3 (12.5%)	5 (21.7%)	0.32
Pulmonary Hypertension	24 (100%)	16 (69.6%)	0.004 *
Highest $PaCO_2$	79.0 [56.3 – 99.3]	57.5 [48.0 – 64.0]	0.006 *
24 Hour SNAP-II Score	32.0 [25.5 – 44.0]	20.0 [6.75 -24.0]	0.002 *
In-Hospital Mortality	**18 (75.0%)** **OR 31.5** [5.6 – 175.9]	2 (8.7%)	<0.001 *

Why should we ever say no?

- Cost
  - Should this ever enter into the decision making process?
  - Must the care team be mindful of costs in the provision of life 'saving' treatment?
  - Are resources truly limited?

Should We Offer ECMO?

Ethics

- Are there ethical implications to offering a life prolonging treatment, without hope for long term survival?
- Does the provision of life prolonging treatment without hope for survival violate 'First, do no harm?'
- Is allowing families to bond with an infant whom is overwhelmingly likely not to survive unethical?

EVOLVE

- NURSING
- PEDIATRIC SURGEON
- NEONATOLOGIST
- RESPIRATORY THERAPIST
- PERFUSION
- PHARMACY
- **MATERNAL FETAL MEDICINE**
- **RADIOLOGY**
- **CARDIOLOGY**

CURRENTLY AVAILABLE TECHNIQUES

- **LHR – Observed/Expected (O/E)**
- **O/E Fetal Lung Volume (MRI/US)**
- Intra-thoracic Liver Herniation
- Percent liver Herniation
- Liver/Thoracic volume ratio
- Stomach location
- Liver/Diaphragm Ratio
- **Pulmonary Artery Dimensions**
  - **The McGoon Index**

Use of magnetic resonance imaging in prenatal prognosis of the fetus with isolated left congenital diaphragmatic hernia. *Victoria T et al. Prenat Diagn. 2012 Aug;32 (8):715-23. Center for Fetal Diagnosis and Treatment, CHOP*

Should we use these criteria?...

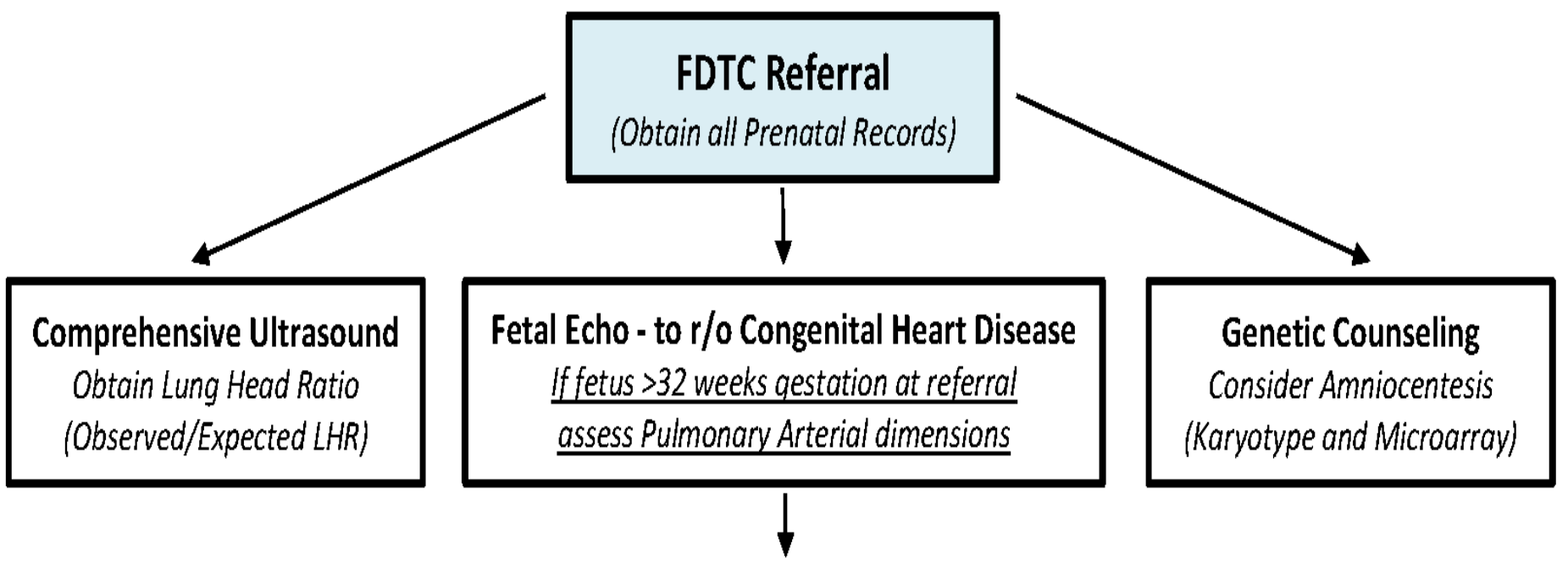
CDH	Parameters	No ECMO	ECMO
Genetics	*Karyotype*	Abnormal	Normal
	Syndrome	Yes	No
Cardiac	*CHD*	Yes	No
	MGI	<1.1	>1.1
Hernia	*Stomach*	Up	Down
	Liver position	Up	Down
Liver	*LiTR*	>14%	<14%
	Percent herniation	>20%	<20%
Lung	*O/E LHR*	<25%	>25%
	O/E TLV	<35%	>35%
	TLV	<15ml	>15ml
	LHR	<1	>1
	PPLV	<15%	>15%

SUMMARY

- What data is predictive for one center may not be predictive for another.
- Specific predictive values needs to be validated internally for each of the center that utilizes imaging technology.
- Combination of measurements give a better prediction of outcome.

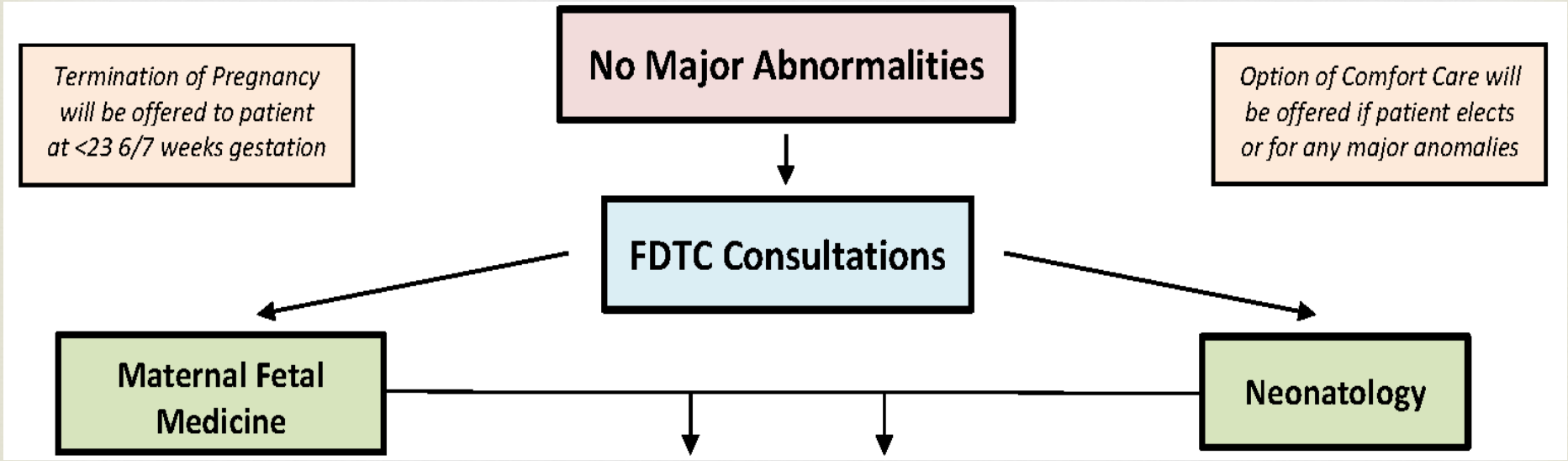
CDH - Prenatal Evaluation Guidelines

UPMC-MWH Fetal Diagnosis Treatment Center (FDTC)



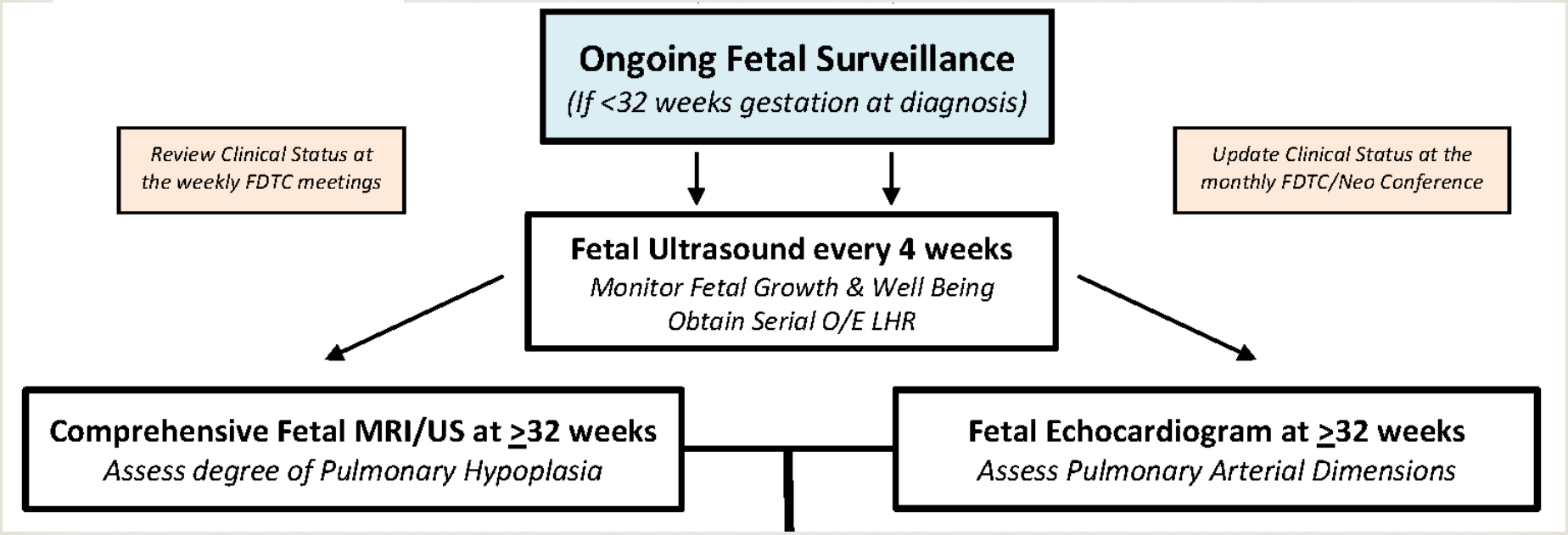
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CDH - Prenatal Evaluation Guidelines

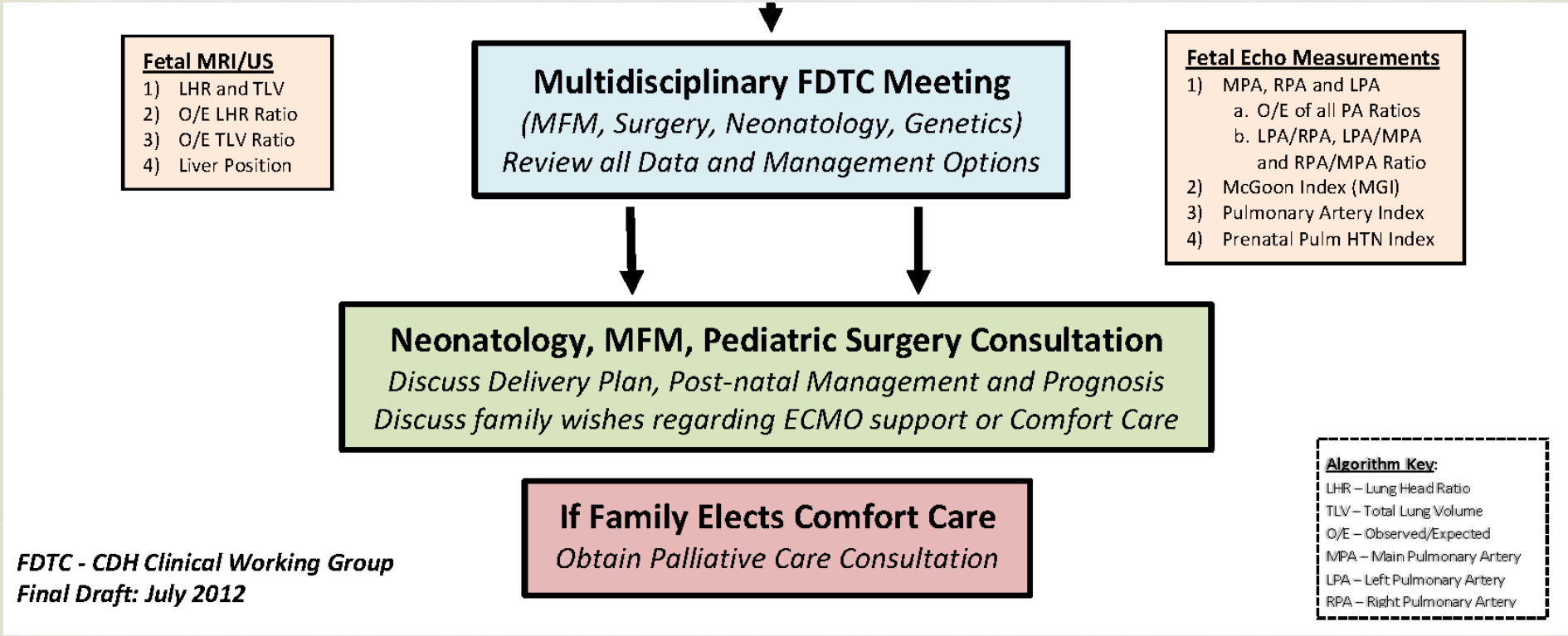
UPMC-MWH Fetal Diagnosis Treatment Center (FDTC)



CDH - Prenatal Evaluation Guidelines

UPMC-MWH Fetal Diagnosis Treatment Center (FDTC)

Review all Data and Arrive to Consensus of Care before Meeting with the Family



FDTC - CDH Clinical Working Group
Final Draft: July 2012

CHP's PROTOCOL

- Delivery Room Airway Management
  - NO bag mask ventilation
  - Intubate immediately
  - Place appropriate size replogle
- Stabilization
  - UMBILICAL LINES
  - Total fluids 80-100 cc/kg
  - Position patient on ipsilateral side

GENTLE VENTILATION

- PIP <24-26
- Rates 40-50
- MAP <16: cautionary 16-22
- Lightly sedated and encourage to breathe

SEDATION

- Avoid use of paralytics
- Gentle sedation as needed
- Morphine boluses or infusion of 0.01mg/kg/hr

PERMISSIVE HYPERCAPNIA

- PCO2 40-60 but tolerate 70-80
- Ph> 7.25

OXYGENATION

- Preductal sats >85
- PaO2> 30
- NO trailed before ECMO for patients who cannot maintain PRE SATS >85, PaO2 >30 or Rising Serum Lactate levels

SUCTIONING

- Q4 hours and prn
- Turn head to ipsilateral side
- Suction only to ¼ cm beyond the ett
- Bagging PIP pressures should be same as ventilator or <26 mmHG
- Allow adequate time for suctioning
- SUCTIOING IS BEST SEDATIVE

HEMODYNAMIC STABILITY

- Normal mean arterial blood pressure for age will be acceptable in most infants.
  - Appropriate range for term infants with CDH is 40-45mmHg
    - **If perfusion and blood pressure do not improve then volume, inotropic support and steroids may be considered**

HYDROCORTISONE

- **Hydrocortisone** is an effective proven therapy for vasopressor and volume resistant hypotension in infants with severe PPHN
- Send a cortisol level at start of therapy; level of <15 considered low
- If the initial cortisol level is low - give Hydrocortisone at a loading stress dose of 2mg/kg/dose
- Start maintenance therapy at 40-50mg/m_2/day (1mg/kg/dose q 8hr)

DOPAMINE

- Usually initiated as the first line inotropic agent
- Start at 5mcg/kg/min; titrate as clinically indicated
- Comfort zone for Dopamine dosing <15mcg/kg/min
- Pulmonary vasoconstriction and reflex tachycardia common at high doses (>15mcg/kg/min)
- Low dose dopamine (3-5mcg/kg/min) can be used to improve renal blood flow via action on the dopaminergic receptors

NITRIC OXIDE

- If patients with CDH have significant pulmonary hypertension than pulmonary vasodilator therapy with inhaled nitric oxide (iNO) may be considered
- The response to iNO in CDH infants is less predictable
- However, iNO may be of benefit in individual cases where ***reactive pulmonary hypertension*** is present
  - Maximum dose of 20ppm
  - If no improvement discontinue

DELAYED SURGERY

- CDH IS NOT A SURGICAL EMERGENCY!

- Advantages of delayed repair:
  - Stabilization of patient
  - Improvement/Resolution of pulmonary hypertension
  - Improve pulmonary compliance
  - Time to evaluate/ manage other abnormalities

ELECTIVE ECMO

- Ultimate protective ventilation strategy
  - Treat reversible PPHN
  - Benefits verses risks
- DOES NOT apply to all CDH patients
  - Pulmonary hypertension verses pulmonary hypoplasia

Where are we now?

- 88 patients (January 2007- current)
- 76 L, 12 R
- 33 ECMO (37.5%)
- Overall survival 67%
  - ECMO 27%, Non ECMO 91%

Abnormal Cranial Exam (30% of survivors)

- Grade I germinal matrix hemorrhage
- Grade II IVH
- Venticulomegaly
- R Venous Sinus Thrombosis
- Hydrocephalus cerebral atrophy

Pulmonary Support
(13.5 % of survivors)

- 1 L NC or less
- NC > 1 L
- Trach/Home vent

Feeding Issues

- 37% of Survivors
- 61% Diagnosis of GERD treated with medication
- 29% with Nissen/GT

What have I learned?

- Despite advances in research, medical management strategies, surgical techniques, and technology over the last twenty years, CDH continues to have a high incidence of morbidity and mortality
- Protocols work and reduce the variability of care amongst different healthcare providers.
- Nurses have a voice!

We cannot save them all but we can find redemption in the fight to try~



- QUESTIONS
- THOUGHTS
- COMMENTS
- CONCERNS