Congenital Diaphragmatic Hernia

What is Congenital Diaphragmatic Hernia?

The diaphragm is a muscle the separates the chest from the abdomen. A congenital diaphragmatic hernia (CDH) happens when this muscle does not close. This allows the organs from the abdomen to move into the chest through this hole. This defect occurs when the baby is developing before birth. It can happen on either the right or left side. In extremely rare cases it can happen on both sides.

The organs that belong in the abdomen move into the chest space and leave less room for the lungs to grow normally. This causes the lungs to be small. After the baby is born there is less blood flow to the lungs and high blood pressure in the blood vessels leading to the lungs. This makes breathing after birth very difficult. It can be life-threatening.

Who gets Congenital Diaphragmatic Hernia?

There is no single known cause of CDH. It tends to happen more in males than females. It is not related to anything a mother did or did not do during the pregnancy. There is also recent evidence that genetics may be related to CDH. Parents who have had one child with CDH are at increased risk to have another child with the same problem.

What are the symptoms?

- Difficulty breathing
- Inability to breathe on their own
- Fast heart rate
- Fast breathing
- Bluish skin color
- Retractions—the muscles between and around the ribs are sucked in
- The abdomen may look flat or sunken in because abdominal organs are in the chest
- The chest may look more full or uneven on each side

What can I expect before surgery?

Doctors and nurses will try to give you a moment to meet your baby for the first time at delivery, but they will be focused on helping him or her in the safest, quickest way possible. Soon after delivery, a doctor will insert a breathing tube (also known as an endotracheal tube) into your baby's mouth and a machine called a ventilator can help your baby breathe. A tube will be inserted into your baby's nose and threaded through the esophagus into the stomach. It will keep your baby's stomach empty so that the baby's lungs have as much room in the chest as possible to expand.

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Once the baby is as stable as possible, NICU physicians, nurses, and respiratory therapists will bring him or her back to our NICU where the baby will remain for the rest of your stay at CHOC.

The diaphragmatic hernia will be surgically repaired once your surgeon and the NICU team feel the baby can safely have the procedure, and they feel the benefits of the surgery are greater than the risks.

After the Surgery:

Your baby will remain on a ventilator until the NICU team decides that he or she can safely breathe on his or her own. During the surgery the abdominal organs were moved from the chest area to the abdomen, so the baby's abdomen may appear full and tight.
Once the team feels the baby is ready, the baby will be fed very small amounts of breastmilk (it is strongly preferred over formula). If the baby is still on a ventilator when it is time to feed, it will be fed through a small tube placed into the stomach through the nose. This is called a nasogastric feeding tube or "NG tube". If the baby is not on a ventilator, we will begin bottle feeding under close supervision of our feeding team. Since your baby's stomach and intestines have never been fed, we watch the baby closely while we start feedings. We will slowly increase the amount as your baby tolerates. IV nutrition will be necessary until your baby reaches a full feeding amount.

What are the long-term concerns? Will this affect growth and development?

Outcomes are usually better in cases where the liver remains down in the abdomen when the defect forms before birth. Babies can have different types of problems requiring long term care and follow up. Many babies will have chronic lung disease and may require oxygen or medications to help them breathe long term. Gastroesophageal reflux is a common problem. This is when acid and fluids from the stomach move out of the stomach and up into the esophagus. This can cause irritation, vomiting, feeding problems, lung problems and growth difficulties. Some babies have developmental delays and may not roll over, sit, crawl, stand or walk at the same time as healthy babies. Developmental therapy is an important part of long term care. Physical therapy, speech therapy and occupational therapy are offered in the hospital and as a part of the discharge plan to help babies achieve their greatest potential.

When should we see a doctor after discharge?

Patients with CDH need long-term follow up with multiple different doctors. Attached is a chart showing the follow up needed for a patient with CDH as they get older. When your baby is discharged you will have the following appointments:

- Pediatrician within 1-3 days after discharge.
- Surgeon 2-3 weeks after discharge from the hospital.
- If there are other specialists providing care for your baby, we will let you know when you will need to have further appointments with them.

You may call to make these appointments around the time of discharge. Some appointments have already been made for you. Your discharge instructions will have the phone number to call with all concerns and questions.

Important Numbers:

CHCO Main Hospital NICU (714) 509-8540 Call for updates on your baby while inpatient

CHOC PSF Surgical Office (714) 364-4050 Call for appointments and concerns after discharge

CHOC PSA Surgical Office (714) 361-4480 Call for appointments and concerns after discharge

	Before Discharge	1-3 Months Of Age	4-6 Months Of Age	9-12 Months Of Age	15-18 Months Of Age	Annually Thru Age 16
Weight, length, head circumference	X	X	X	X	X	X
Chest X-ray	X	If patch present	If patch present	If patch present	If patch present	If patch present
Pulmonary function tests **(Pulmonary consult and long-term f/up)**			If indicated		If indicated	If indicated
Childhood Immunizations	X	X	X	X	X	X
RSV Prophylaxis **(RSV Clinic CHOC)**	RSV season during 1st 2 yrs of life (during RSV season the first dose is given prior to discharge or on the day of discharge)					
Echocardiogram and cardiology follow-up	X	If previously abnormal or on supplemental oxygen	If previously abnormal or on supplemental oxygen	If previously abnormal or on supplemental oxygen	If previously abnormal or on supplemental oxygen	If previously abnormal or on supplemental oxygen
MRI	Head CT or MRI if there was an abnormal finding on head ultrasound; or seizures/abnormal neurologic findings; or ECMO or patch repair	As indicated	As indicated	As indicated	As indicated	As indicated
Hearing evaluation	X	X	X	X	X	Every 6 mo to age 3, then annually to age 5
Developmental/ Neurodevelopmental screening evaluation **(EDAC)**	X	X	X	X		Annually to age 5
Assessment for oral feeding difficulties **(outpatient speech therapy)**	X	X	If indicated	If indicated	If indicated	If indicated
Upper GI and/or pH probe **(GI Consult for reflux management)**	Consider at any time/ age for all patients if symptomatic					
Scoliosis and chest wall deformity screening (physical exam, chest X-ray, chest CT) **(Pediatric Surgery and Pediatrician f/up)**				X		X
Pediatric Surgery Outpatient Follow-up	First outpatient visit scheduled for 2-3 weeks after discharge	X	X	X	X	X