High airway obstruction

Patient Information Series – What vou should know. what vou should ask.

What is high airway obstruction?

Congenital High Airway Obstruction, (sometime called “CHAOS”) is a very rare condition whereby the upper airways of the fetus can be completely blocked, resulting in trapping of lung fluid in the lungs of the developing infant and poor development of the underlying lung tissue before birth.

How common is this obstruction?

Very few cases of this condition have been reported in the medical literature making it an exceedingly rare condition.

What causes Congenital High Airway Obstruction?

Sometimes underlying structural problems in the lungs or surrounding tissues can cause compression of the airways leading to this condition. In other cases, an inherent problem during fetal development of the airways when the organs are forming early in pregnancy may cause the problem.

How is this problem diagnosed?

Usually at the time of routine ultrasound to look at the baby’s anatomy, this problem can be found incidentally. This condition will be suspected if the ultrasound shows enlarged and very bright lungs on the scan, flattening of the diaphragm, a shirt in heart placement towards the middle of the chest and/or fluid filled airways below the obstruction. On some occasions, signs of heart failure may be present with fluid around the heart or lungs or in the abdomen of the fetus or an unusually large amount of amniotic fluid surrounding the fetus.

Are there other problems associated with this condition?

Sometimes this condition can be associated with a condition called Fraser Syndrome, a complex genetic problem resulting in structural differences in the eyes (fused eyelids), digits (fused fingers or toes), genitalia or urinary tract. This is caused by a mutation in one of several responsible genes.

Should I have any special testing performed if my baby has this condition?

Once diagnosed, other tests such as genetic amniocentesis, specialized fetal heart ultrasound fetal echo) and detailed ultrasounds will be offered to rule out other structural problems. Consultations with other health professionals such as fetal medicine specialists, neonatologists and Pediatric/ ENT surgeons will be advised.

What is the prognosis for the baby after birth?

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This is a serious condition that is often fatal. If no other structural or genetic differences are present in the fetus, the outcome will depend on the lung growth and whether the baby develops signs of heart failure (fetal hydrops). Rarely there can be spontaneous resolution of the obstruction in fetal life.

What can be done to treat the fetus in this condition?

There have been early reports of some fetuses responding to fetal surgery whereby a scope is passed through the windpipe of the fetus in an attempt to bypass the obstruction and decompress the lungs. This approach may not be applicable to all fetuses with this condition however, based on the severity of the obstruction and the gestational age of the affected fetus. Due to the severity of the condition, termination of pregnancy is also an option that is discussed with parents.

What happens at delivery of affected infants?

These infants can have immediate problems impairing their ability to breathe at birth. Therefore a special delivery procedure, called an “EXIT” procedure may be arranged. In this procedure, once the baby’s head is delivered, delivery of the rest of the baby is deferred until an airway can be secured by a specialist physician or surgeon highly trained in airway management of infants. These infants need to be cared for in high-level neonatal intensive care units and may have significant medical and respiratory problems in their first year of life requiring prolonged hospitalization. Longer term developmental problems in childhood have also been reported in survivors with this condition.

Will this problem recur in subsequent pregnancy?

Often this problem is a sporadic one with low chance of recurrence. If it is associated with a genetic mutation leading to a recognizable syndrome such as Fraser Syndrome there is a 25% chance of having another affected fetus.

What other questions should I ask?

- Is this a lethal problem for my baby?
- Are there any other structural problems in the fetus?
- Should I have genetic testing?
- Is my baby a candidate for surgery before birth?
- Where is my baby most safely delivered?

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