Binder syndrome (nasomaxillary hypoplasia)

Patient Information Series – What you should know, what you should ask.

What is Binder Syndrome?

Binder syndrome is a problem of part of the nose and upper jaw. Affected individuals have a very flat nose and an underdeveloped upper jaw. The characteristics of the syndrome are typically visible when looking at the profile of the face. The profile looks flat with a relatively protruding lower jaw.

The facial anomalies in Binder syndrome may be isolated. Possible associated anomalies are seen in the bones (especially short upper arms and upper legs and neck anomalies), hearing difficulties eye movement disorders, thin lining of the inner part of the nose, small frontal sinuses and small upper front teeth. There is no proof that Binder's syndrome is associated with lower intelligence.

How does this syndrome happen?

Binder syndrome is a rare problem and the causes are unclear. Mothers with autoimmune diseases, a lack of vitamin K, alcohol abuse or mothers using warfarin (a certain form of blood thinner) or phenytoin (a medication to control seizures) have a higher risk in a baby with Binder syndrome. However, most cases have not been found to be related to any of these factors and happen by bad luck.

Should I have more tests done?

You should ask if a specialized/advanced ultrasound of the baby during the pregnancy can be done to see if the baby shows signs of other problems. Sometimes MRI (a different way to obtain images of a fetus inside the womb) can be performed to detect problems of the neck. Some problems seen in combination with Binder Syndrome cannot be seen before the baby is born, such as changes in the front teeth or hearing difficulties. There is no genetic test available to prove that the baby has Binder syndrome.

Binder syndrome may be part of another syndrome, which may be more complicated. When other anomalies are visible the risk that Binder syndrome is part of another syndrome is high. In this situation, it is important to have a consultation with a genetic specialist. It is possible that invasive testing such as amniocentesis (where a sample of the water around the baby is collected with a small needle) will be offered to you to detect or exclude some of these syndromes. Amniocentesis carries a small risk of miscarriage, and some women choose not to have it for fear of losing their baby. Not all syndromes can be diagnosed by an amniocentesis and not all anomalies are visible on ultrasound examination. However, when there are no other anomalies visible on ultrasound and the amniocentesis gives normal results, Binder syndrome is very likely to be isolated.

What are the things to watch during the pregnancy?

Sometimes babies with Binder syndrome will accumulate extra water (also called amniotic fluid) around themselves during the pregnancy, usually because the baby has problems swallowing the amniotic fluid due to the problems with the nose and mouth. This condition is called polyhydramnios. It can stretch the uterus too much and cause early labour well before the due date. Knowing this in advance, your doctor or midwife can help with decreasing the risk of an early birth. That is why most specialists will recommend regular ultrasound examinations at least every 4 weeks.

What does it mean for my baby after it is born?

Babies born with isolated Binder syndrome usually do well. Some babies experience breathing or drinking problems immediately after birth because the nose is too small. These babies may need oxygen or the placement of tubes to assist breathing. It is advisable to discuss the best place of delivery with your caregiver.

Many children who have a severe form of Binder syndrome will undergo plastic surgery for cosmetic reasons or dental treatment later in life. When Binder syndrome is part of another syndrome, the baby may have more problems after birth. Then, how the baby does is much more variable and depends on the kind and severity of the underlying problem.

Will it happen again?

The risk that isolated Binder syndrome will happen again is very low. If Binder syndrome is part of another syndrome, then the risk that it will happen again depends on the type of underlying condition and a consultation with a genetic specialist may be helpful to help sorting this out.

What other questions should I ask?

- Does this look like a severe form of Binder syndrome?
- Are there other anomalies visible?
- Is invasive testing recommended?
- How much is the amount of amniotic fluid?
- How often will I have ultrasound examinations done?
- Where should I deliver?
- Where will the baby receive the best care after it is born?
- Can I meet in advance the team of doctors that will be looking after my baby when it is born?

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