Bifid Nose

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

What is a bifid nose?

A bifid nose is a rare birth defect. The nose appears divided into two halves. There is a large degree of variability, ranging from a small hardly noticeable groove at the tip of the nose to a complete division of the nose. A bifid nose is often associated with wide-spaced eyes (hypertelorism), an opening in the lip and roof of the mouth (cleft lip and palate), or a skin-covered gap in the skull. (encephalocele)

How does a bifid nose happen?

A bifid nose is caused by the failure of the two sides of the nose to fuse during the first two months of development in the womb. In some cases, a bifid nose is the only defect in an otherwise healthy baby. This may happen by chance through a local problem during the development of the nose or maybe the result of a genetic change passed down from one or both parents.

Sometimes, a bifid nose is part of a syndrome. A syndrome is a set of symptoms, signs, or defects that together are characteristic of a specific disorder. A syndrome often associated with a bifid nose is BNAR syndrome. In addition to a bifid nose, people with BNAR syndrome can have kidney, anus, and rectum defects. The role of genetics in being born with a bifid nose is not completely understood yet.

Should I have more tests done?

You should ask if a specialised/advanced ultrasound of the baby during the pregnancy can be done to detect other anomalies. Sometimes MRI (a different way to obtain images of a fetus inside the womb) can be performed.

When other anomalies are visible the risk that the bifid nose is part of a syndrome is high. In this situation, it is important to have a consultation with a genetic specialist. It may be that invasive testing (amniotic fluid puncture) 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 genetic investigations such as an amniocentesis and not all anomalies are visible on ultrasound examination. However, when there are no other anomalies visible on ultrasound and invasive testing gives normal results the bifid nose is very likely to be isolated.

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What are the things to watch for during pregnancy?

If there are no other anomalies, then you can have your pregnancy checks as normally planned. If there are other anomalies, changes in your care will depend on what they are and what they mean. It is advisable to discuss the place of delivery with your caregiver.

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

A baby with a bifid nose usually needs reconstruction surgery, which can be challenging for severe cases. Usually, a specialised craniofacial team is involved. This team will determine the appropriate treatment and follow-up plan. The prognosis is variable and also depends on the kind and severity of possible other anomalies.

Will it happen again?

The risk that an isolated bifid nose will happen again is very low. If the bifid nose is caused by a genetic change or is part of a syndrome, then the risk that it will happen again depends on the type of genetic change or underlying condition. A consultation with a genetic specialist may be helpful to sort this out as there are several different patterns of inheritance for a bifid nose.

What other questions should I ask?

- Does this look like a severe form of bifid nose?
- Are there other anomalies visible?
- Is invasive testing recommended?
- 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?

Last updated November 2022