

# Nose Proboscis

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

## **What is Proboscis?**

A proboscis is a developmental problem of the nose. The nose or half of the nose is replaced by a poorly developed, tubular, nose-like appendage.

Most proboscises are located in the midline of the face. Very rare cases are located next to the midline. These are two very different disorders.

### **Midline proboscis:**

A midline proboscis is located in the midline of the face, above or between the eyes. A midline proboscis is always associated with an absent nose, severe eye anomalies (sometimes there is just one central eye) and very severe brain anomalies (named holoprosencephaly). These severe forms of holoprosencephaly combined with proboscis are often associated with chromosome abnormalities. Chromosomes are where most of our genetic information is kept. We usually have 46 of them matched in pairs: 23 come from one parent and the other 23 come from the other parent. For example, people with Trisomy 13 have an extra chromosome number 13. That condition is often seen in babies with a midline proboscis: if such a change in information within the chromosomes occurs, it would be the cause for the proboscis. Babies with Trisomy 13 sadly usually die during the pregnancy or soon after being born.

### **Lateral proboscis:**

In very rare cases the proboscis is located next to a normal nose or next to a half nose. This is named a lateral proboscis. A lateral proboscis is a different kind of proboscis than a midline proboscis. A lateral proboscis can be an isolated anomaly but the risk on eye anomalies or clefts in the upper lip, palate or the cheek is high. Brain anomalies are sometimes present but rare. Intelligence is usually normal.

## **How does a lateral proboscis happen?**

It is not clear why a lateral proboscis develops next to a normal or half nose.

## **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. Special attention should be paid to the brain, eyes, upper lip, palate and cheeks as anomalies of these areas are associated with this type of proboscis. Sometimes, MRI (a different way to obtain images of a fetus inside the womb) can be performed to detect brain or skull anomalies.

## **What are the things to watch for during the pregnancy?**

When there are no other anomalies, you can continue to have your regular pregnancy check-ups. If other anomalies exist, they may lead to changes in your pregnancy care.

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

The prognosis of babies born with this type of lateral proboscis and without other major anomalies is generally good. Normal developmental outcome is the rule, unless brain abnormalities are identified. Breathing, eating or drinking problems are not commonly mentioned, but may occur. It may therefore be advisable to discuss the place of delivery with your caregiver based on the appearance of the face of the baby.

Plastic and reconstructive surgery is required to correct this problem. Given the high degree of variability in the appearance of the lateral proboscis, an approach specific to your baby will be required. Final cosmetic reconstruction is usually performed after facial growth has been completed; achieving an aesthetic result is often a challenge. Aesthetic and psychological problems need attention and a multispecialty team is usually necessary to evaluate, discuss and treat possible physical or emotional problems that comes with anomalies of the face.

## Will it happen again?

The risk that a lateral proboscis will happen again is very low. The risk is higher when parents are related to each other.

## What other questions should I ask?

- Does this look like a severe form of proboscis?
- Are there other anomalies visible?
- 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 (especially a plastic surgeon) that will be looking after my baby when it is born?

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