What is Pierre-Robin-Sequence?

Pierre-Robin-Sequence (PRS) is a congenital condition, meaning it is present before birth. The most prominent symptom is trouble breathing due to airway obstruction.

This is caused by

- mandibular retrognathia (abnormal positioning of the lower jaw), sometimes also micrognathia (unusually small lower jaw)
- Glossoptosis (obstruction of the upper airway caused by the tongue base falling backwards).

The majority of newborns with PRS also have a cleft palate (incomplete closure of the roof of the mouth, commonly U-shaped).

What causes PRS?

PRS can be caused by mechanical stress in the womb (non-syndromic PRS) or be associated with other conditions (syndromic PRS) such as Stickler-Syndrom, DiGeorge-Syndrom (=Velocardiofacial Syndrome; VCFS) or Treacher-Collins-Syndrome to name but a few.

How can PRS be diagnosed?

PRS can be diagnosed by an ultrasound exam before birth. The diagnosis is then confirmed by clinical features found in the regular postnatal exam shortly after birth. If there are no breathing difficulties present, it is not PRS.

How can PRS be treated?

Primary Treatment

There is a serious risk of damage to the newborn during delivery. Therefore, it is mandatory to build up an interdisciplinary team including an obstetrician, neonatologist, pediatrician, anaesthesiologist / anaesthetist and a maxillofacial surgeon who are present at the time and after delivery. However, this requires a hospital where all these services can be provided.

Secondary Treatment

In principle, there are two alternative treatment options, surgical or conservative. However, although the surgical approach is more common, we prefer the conservative approach.

Conservative treatment:

At our clinic we prefer the conservative option which is not invasive (no cuts, no stiches, no anaesthesia), thereby reducing stress on the newborn considerably. The method is scientifically well proven and safe. We have treated a great number of newborns with PRS and not in a single case a surgical intervention was necessary.

After a short exam to determine the local situation, a plate is manufactured and individually adjusted to the palate and tongue base. It gently pushes the tongue base forward, immediately preventing airway obstruction, hypoxia and asphyxia. The baby can instantly breathe and feed normally. Regular control exams to be sure the plate sits correctly are conducted on an ambulatory basis. Over time, usually 2-3 months, the device induces mandibular growth, the jaw grows larger, until a stable situation is established. The plate is no longer needed and can finally be removed.

Surgical treatment:

There are several surgical methods which are all invasive:

1. Glossopexia: Fixation of the tongue to the lower lip by stitches;
2. Distraction osteogenesis: Cutting the mandible and enhancing growth by a mechanical device which is inserted onto the bone and activated by daily turning a screw poking through the skin
3. Tracheotomy: Making an incision on the frontal aspect of the neck and forming a canal in order to insert a tube into the windpipe.

Closure of the soft and hard palate:

The cleft palate is addressed later (age 4-6 months and 11 ib. / 5 kg weight) in a separate operation.

How is the prognosis?

When treated at an early stage, i. e. during the first days after birth, children reach full development and size. They usually grow to lead a normal and healthy life. However, if treatment is delayed, children might grow below average size and show developmental insufficiency. This is due to early chronic hypoxia as well as lack of nutrition due to feeding problems as well as associated stress.