

## Pierre Robin Syndrome (PRS)

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Congenital condition named after French doctor who identified its main characteristic of this syndrome featured as small lower jaw (Micrognathia) with receding chin with glossoptosis (Tongue falls backwards into pharynx and throat). Usually the tongue appears large due to under develop mandible and floor of the mouth.

Poswillo (1966), mentioned the cause of this syndrome is due to postural intra uterine defect associated with other postural defects such as congenital hip dislocation and talipes. Cleft of palate occur due to large tongue thrust between the two palatal shelves, the tongue frequently wedged in the characteristic U shaped cleft of the palate in the neonatal period leading to respiratory crisis, asphyxia and inspiration pneumonia due to airway obstruction after birth.

Recent studies revealed a genetic cause to Pierre Robin Syndrome was identified and caused by genetic anomalies of chromosome 2, 11 or 17 with changes in the DNA near and the SOX9 gene are the most cause of isolated cause of PRS.

Some people have the features of Pierre Robin Sequence as part of syndrome that effect other organs and tissue in the body such as Sticker Syndrome or Campo melic dysplasia syndrome.

The name of Pierre Robin Syndrome replaced as Pierre Robin Sequence, because one of its features of underdeveloped mandible sets of sequence of events before birth that causes other signs and symptoms, that small lower jaw and under develop floor of the mouth effects displacement of the large tongue backwards blocking the airway and thrusting the tongue between the two palatal shelves and preventing the fusion of the two palatal shelves greatly effect of formation of palate during embryonic development before birth.

These changes in the lower jaw with large tongue and cleft palate, these occurred during embryonic stage and after birth greatly effecting feeding of new borne baby and he did not gain weight and growth.

The incidence of Pierre Robin Sequence is quiet rare and the American studies showed 1/14000 babies born with Pierre Robin sequence.

From our long experience, we did consider the new born baby with Pierre Robin Sequence featured glossoptosis, micrognathia, cleft palate with obstruction of airway and grasping air with body looks cyanosed an urgent and emergency treatment required.

Our technique is by using an immediate glossy-boxy technique by inserting a straight needle carrying black silk suture size 0, the needle passed between the posterior third of the tongue and anterior 2/3<sup>rd</sup> of the tongue with slight traction and sutured to the lower alveolar ridge of the mandible to prevent backward movement of the tongue and making the anterior part of the tongue with free movement and a feeding aid by using an acrylic plate to cover the palate and cleft to help the baby in breast feeding by holding the breast nipple under Neath against the acrylic feeding aid in the palate for creating negative pressure for milking the nipple of breast.

The baby should lie on his abdomen with a pillow under the shoulder without touching or any pressure on lower jaw of the baby. by this technique we gain a remarkable growth of the mandible and floor of the mouth within 3 - 4 weeks, this technique is a lifesaving technique.

Microsomia or small mouth opening usually associated with Pierre Robin Sequence, we advise the mother of the baby to practice daily stretching of the corners of the mouth on both sides in the same time by gentle movement with steroid cream of 1% to improve the mouth opening within few months.

Other deformities may not correct till the age of 2 - 3 years for cleft palate. Jaw deformity by distraction technique during growth period of the child while other deformities' might be delayed until the adolescence when growth completed.

We did an experimental research studies on Rabbits for elongation of lower jaw of the Rabbit by distraction technique. Distraction achieved by three phases, surgical phase, latent period phase and consolidation phase. The most critical and vital point of distraction is the latent period phase which elapsed between 3 - 7 days.

We did found by our research and by studding the latent period phase after surgical phase a gap created by osteotomized of the bone. During latent phase a healthy granulation tissue formed with mesenchymal stem cells derived from the bone marrow and periosteum with heavy formation of fibroblasts with growth factor effect and the fibroblasts oriented with the direction of distraction force with new bone formation by stretching bone by rhythmic distraction technique of 1 mm/day divided in 2 terms of 0.5 mm, we did achieve 10 mm elongation of Rabbit mandible.

At the end we believe experimental surgery on good animal model achieve great benefit for humanity and to improve and advance surgical field of Maxillofacial Surgery.

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