PIERRE ROBIN SYNDROME*

ANANTA BHOGAONKER, KIRAN BALA SAGAR AND O. N. BHAKOO

Report of a Case

Chandigarh

The Pierre-Robin syndrome consists essentially of micrognathia and the consequent glossoptosis. Cleft palate is seen in a little over half of such patients and occasionally, a high-arch may be the only palatal abnormality. It is one of the important causes of dyspnoea and cyanosis in the neonatal period (Davis and Dunn 1933).

We record below a typical case of Pierre Robin syndrome and discuss simple, yet effective, measures for management of such a case.

Report of a Case

Baby, a 4-day old male infant was brought to our hospital with the history of feeding difficulty, nasal regurgitation of feeds, dyspnoea and attacks of cyanosis since birth. The baby had been delivered at full term to a 25-year old mother. He cried soon after birth and a few minutes later became cyanosed. The attending doctor resuscitated him. He was fed by means of a small pipette as he was unable to suck. However, feeding was often associated with regurgitation of milk, dyspnoea and cyanosis. On the 4th day of life, the child developed marked dyspnoea and cyanosis.

The course of pregnancy had been uneventful. Family history revealed that one maternal cousin had a cleft-lip and cleft-palate.

On physical examination, the child weighed 2.5 Kg., the height was 21”, skull circumference 13”, temperature 98°F and respiratory rate 64/minute. The baby had mild physiological jaundice, cyanosis, micrognathia, incomplete cleft palate and glossoptosis. The conjunctivae were inflamed and the anterior fontanelle was normal. The eyes including the fundii and ears were normal. Moro’s reflex was absent while the sucking reflex was present. The chest showed a few scattered crepitations at both bases. The heart was clinically normal.

The child was nursed in a prone position with a small pillow under the chest and oxygen was given. Crystalline penicillin 50,000 units 12 hourly was administered ever 5 days. To avoid recurring attacks of cyanosis due to air-way obstruction, a plastic air-way was used. Expressed breast milk was given by gavage and he was

*From the Pediatric Unit, Institute of Post-Graduate Medical Education and Research, Chandigarh.
gradually trained for a bottle-feeding in the upright position. He did not require the artificial air-way after the 12th day of hospitalisation. He started gaining weight from the 5th day of hospitalisation and was discharged after the mother had learnt the feeding technique and postural management of the infant.

Discussion

Though the syndrome is named after Pierre Robin because of his extensive reviews of the condition, it had been described earlier by Shukowsky in 1911. In this symptom-complex the child has an “Andy-Gump or bird face” appearance because of micrognathia. This is the determining cause of glossoptosis. The genioglossus is attached to the symphysis menti and is unable to exert its full forward pull because of the hypoplastic mandible. Consequently, due to poor anchorage the tongue falls back into the hypopharynx, presses on the epiglottis and acts as a ball-valve, letting the air out but preventing its entry. As a result, cyanosis, dyspnoea, substernal retraction and poor feeding follow, leading to the so-called glossoptotic cachexia. Aspiration pneumonia is another hazard and if diagnosed late or inadequately treated may prove fatal.

Some other features which have been recorded in association with this condition are kyphosis, genu valgum, strabismus, pigeon breast and flat foot, Pierre Robin thought that these result from pre-existing glossoptosis and secondary respiratory and swallowing difficulties (Eley and Faiber 1930). It may also be associated with many other features like ankyloglossia (May and Chun 1948), heart lesions like patent ductus arteriosus, atrial septal defect, coarctation of aorta, anomalies e.g., cataracts, glaucoma, retinal detachment, Moebius syndrome, coloboma of the choroid, (Smith et al. 1960) etc. Auditory defects like deafness, otitis media and low-set ears have been noted by Smith and Stowe (1961). Congenital multiple arthrogryposis, congenital arthro-myodysplasia and foetal chondrodysplasia are known to co-exist in this syndrome. None of these congenital abnormalities however, were seen in our case.

Management

The principles of management pivot round the basic abnormality glossoptosis, which causes most of the mechanical problems in breathing and swallowing. The subject was recently reviewed by Goldberg (1962.)

The immediate supportive treatment is to put the child in a prone position with the face down and with the chest elevated on a small pillow. This is known as “orthostatic nursing.” During a bout of crying, it is important to hold the baby on its stomach in order to prevent glossoptosis and respiratory embarrassment.

The baby, and more essentially the mother, need to be trained for orthostatic feeding. In this technique, the baby sucks with the thorax held upright so that he lifts his neck and propels his chin forward like the young ones of other mammals. The mother may have to place the nipple in the infant’s mouth and help him to hold it, but one must avoid any pressure on the chin of the child. He should be held erect and be carried...