Converse: Chapter 50

The Robin Anomalad: Micrognathia and Glossoptosis With Airway Obstruction

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When micrognathia (small jaw) and glossoptosis (falling backward of the tongue) occur in the newborn, there is a great danger of upper airway obstruction. These deformities are frequently associated with an incomplete cleft of the palate, and the entity has been referred to as the Pierre Robin syndrome. Robin was far from the first to recognize the anatomical condition that bears his name, but he does deserve credit for calling attention to the clinical significance of this problem. He pointed out that in severe cases death was inevitable. In spite of his publications, it has taken many years for this knowledge to become widespread, and for an effective method of treatment to be developed (Robin, 1923a, 1927b, 1928, 1934).

Gorlin, Pindborg, and Cohen (1976) noted that the condition is not a genetic "syndrome" and prefer the term "Pierre Robin anomalad". An anomalad is described as "a malformation together with its subsequent derived structural changes".*

* "Anomalad" was coined to meet the need for a term referring to a group of anomalies rising from the same developmental defect. It is used to refer to a group of anomalies in the sense that triad refers to a group of three, and tetrad to a group of four. - F. Clarke Fraser.

Definition

Micrognathia is the term most frequently used to describe the anomaly of the jaw, although many other terms have appeared in the literature, such as hypoplasia of the mandible, mandibular hypotrophy, congenital mandibular atresia, brachygnathia, ateliosis of the mandible, and hypomicrognathia (Lenstrup, 1925; Eley and Farber, 1930; Robin, 1934; Callister, 1937; Weisengreen, 1940; Schwartz, 1940; Llewellyn and Biggs, 1943; Longmire and Sanford, 1949; Beers and Pruzansky, 1955; Benavent and Ramos-Oller, 1958; Oeconomopoulos, 1060).

Retrognathia is a more accurate and more inclusive term inasmuch as several conditions can lead to a posterior displacement of the chin. The retroposition of the chin is a frequent finding common in many types of jaw deformities, and when retrognathia is associated with glossoptosis and respiratory obstruction, it fits Pierre Robin's original description.

Retrognathia can be subdivided into several types of anomalies, including those with a normal mandibular size at birth and a posterior displacement of the chin by external pressure. Such a mandible would have an excellent growth potential and is probably the type most frequently seen in the Pierre Robin syndrome (the Robin anomalad). In addition, there are some mandible which have a definitely reduced size and diminished growth pattern, and other cases in which this abnormal mandibular growth pattern is associated with diminished growth potentials of the adjacent facial bones and distortion of the cranial base as well.
However, because the term micrognathia is so widely used, it will be employed in this chapter.

**Historical Aspects**

In 1822, the deformity of micrognathia was reported by Saint-Hilaire, and in 1891 Taruffi subdivided the defect into two categories - hypomicrognathus (small jaw) and hypoagnathus (absence of jaw). In 1891, Lanelongue and Manard reported four patients, two of whom had cleft palates (Eley and Farber, 1930). In 1902, Shukowsky described a surgical adhesion of the tongue to the lip to correct the condition, but the usefulness of this operation was not widely appreciated (Shukowsky, 1911). A further description of this mandibular problem was published in 1922 by Gladstone and Wakely.

In 1923, a Parisian stomatologist named Pierre Robin published a paper calling attention to the problems associated with glossoptosis and obstruction of the airway (Robin, 1923a). He said that they were caused by hypoplasia of the mandible and that in 1902 he had first described a prosthetic appliance which he called a "monobloc" and which he used to restore the relationship between the upper and lower jaws. Robin continued to write profusely on the problems of glossoptosis and "mandibular hypotrophy". He pointed out that glossoptosis in infants could cause episodes of cyanosis and predispose to any of a number of pulmonary complications in the newborn. He further stated, "I have never seen babies live for more than 16 to 18 months who presented hypoplasia such that the lower maxilla was pushed more than 1 cm behind the upper" (Robin, 1934).

He also described in detail the feeding problems of these children and their failure to gain weight which often was serious and led to death from athrepsia. Robin, however, became quite carried away by the problem of glossoptosis, its prevalence and the dire effects that it produced. He thought that it was present in three out of five children and that, if not corrected, it predisposed to "protruding ears, kyphosis, scoliosis, lordosis, strabismus, adenoids, carious teeth, flat feet, harelip, rachitic rosary, cryptorchidism, ... appendicitis, constipation and enuresis (Robin, 1929b; Schwartz, 1940; Randall, Krogman and Jahina, 1965). Cleft palate is described in one of his cases.

In 1930, Eley and Farber described three cases and pointed out that micrognathia was an important cause of cyanosis during infancy and one which at that time was seldom included in the differential diagnosis of this condition. They treated their patients with a head apparatus designed to force the jaw forward. Since that time, many methods of treatment have been described, thus leading to the surgical procedures that are favored today. Perhaps the most significant of these developments was the contribution of Douglas (1946), who showed that many lives could be saved by suturing the tongue forward to the lower lip. Through his personal cases and information obtained from an extensive poll of plastic surgeons, he demonstrated an appreciable decrease in the death rate among these infants when the problem was recognized and surgical treatment instituted (Douglas, 1946, 1950, 1956).

**Etiology**

The exact cause of retrognathia is not known, and since several different anatomical conditions have been described as being associated with it, it seems likely that there is no
single etiologic factor. The possibility that it is inherited has been proposed, and individual case histories have shown a familial tendency in some instances (Kiebel and Mall, 1910; La Page, 1937; Fraser and Calnan, 1961). However, in larger series there has been little evidence of any genetic influence (Gladstone and Wakely, 1923; Bromberg and associates, 1961).

Chapple (1955) pointed out that "the structures involved arise from different anlagen, from different layers at different times. A gene covering this odd assortment has no parallel. The structures are in no other way related, to our knowledge, but by their mechanical relationship where the one can so readily affect the other".

There seems to be good evidence that intrauterine pressure may well be the most frequent causative factor. Sharp flexion of the head downward and forward would place the chin up behind the manubrium and prevent the jaw from developing forward. Davis and Dunn (1933) pointed out that the mandibular distortion was probably due more to forcing the mandibular angle from an obtuse angle to a right angle than to an underdevelopment of the bone.

Parmelee in 1931 described a number of deformities in infants that could be explained by an abnormal intrauterine position. Such a position can be determined shortly after birth by "folding up" the head and extremities until a "position of comfort" is found. In a baby born by breech delivery, the position of comfort is noted when the legs are sharply flexed at the hips. When they are extended, the baby is very uncomfortable.

Chapple (1941, 1955) related these data to the problem of micrognathia and found a good parallel. He further pointed out that, until six weeks of age, the conformation of the embryo is nearly circular. Then the head begins to rise from the cephalic flexion: "In the early life of the embryo, while his head is still on his chest, the tongue lies between the sides of the palatal arch. His head must be lifted from his chest to allow the tongue to fall down into the mouth so that the sides are free to unite into an arch. If it is not raised, the already intact tongue prevents their union, and the tongue grows into the nasopharynx". This would explain the high incidence of cleft palate (Eley and Farber, 1930; Chapple and Davidson, 1941; Chapple, 1950, 1955; Bigotti, 1959). The flexed position produces all the abnormalities seen in the Pierre Robin anomalad, and if the head is maintained in this position past the usual time, these abnormalities are likely to persist to a greater or lesser degree.

Cleft palate is frequently seen with micrognathia, cleft lip very rarely (Kiskadden and Dietrich, 1953; Benavent and Ramos-Oller, 1958; Bromberg and associates, 1961). Kiebel and Mall (1910) considered the fourth and fifth months of pregnancy the critical period for the occurrence of the deformity. A variation in the timing might well produce a cleft palate in some and not in others. Davis and Dunn (1933), Llewellyn and Biggs (1943) and Sweet and Kemsley (1947) all were of the opinion that the weight of the baby on the facial structures in utero must have some bearing on the development of the condition.

Because many of these patients eventually show remarkable mandibular growth and reach normal mandibular proportions, it seems likely that external pressure on normal structures could account for this sequence of events (Walker, 1961). The mandibles of other patients remain hypoplastic, and it may be that in these patients an intrinsic factor causing a reduced mandibular size produces the deformity. Still other children can be shown to have
underdevelopment of adjacent facial bones and distortion of the cranial base in addition to the underdevelopment of the mandible.

In summary, retrognathia with glossoptosis seems to be caused by one of several factors: (1) normal mandibular facial potential but intrauterine inhibition probably due to external pressure; (2) local failure of mandibular growth with normal facial bone development; or (3) mandibular failure associated with other craniofacial abnormalities causing a regional failure of growth (Krogman, 1962; Randall, Krogman and Jahina, 1964).

Robin wrote as follows: "Mandibular hypotrophy is never idiopathic. As a rule it is caused by congenital syphilis or tuberculosis, by hereditary dystrophia from alcoholism or by some other infection. Occasionally, a mild case occurs in the child of parents, one of whom has large jaws and the other, narrow ones: the child has a narrow upper jaw and a broad lower one or vice versa, so that there is a lack of equilibrium between the two and functional troubles appear" (Robin, 1934).

Diagnosis

The child with micrognathia has a characteristic appearance, with an undershot chin. When seen, he should arouse immediate suspicion of the likelihood of partial airway obstruction, feeding difficulties, and a possible cleft palate. There will be variations in the degree of retrognathia, the severity of the airway obstruction, the feeding difficulties, and the age at which these problems arise.

Respiratory Obstruction. The difficulty in breathing is caused by the glossoptosis, or the impingement of the base of the tongue on the posterior pharyngeal wall and pressure on the epiglottis, causing a ball-valve type of obstruction of the glottis and inspiratory obstruction. Typical retraction of the intercostal spaces, the suprasternal space and the epigastrium is seen along with activity of the accessory respiratory muscles. In addition, there is an obstructed sound to the baby's breathing, and there may be a history of choking and cyanotic attacks. These findings in the newborn should make one suspect micrognathia. The deceptive aspect of the respiratory obstruction is that these children can frequently maintain an adequate airway if they struggle, strain, and cry. They may not even appear to suffer from obstruction, and the diagnosis of airway obstruction may be missed. When the infant relaxes or dozes, obstruction occurs, he awakens and cries again. As a result, he can literally exhaust himself to death unless the obstruction is relieved. Delay in treatment can cause brain injury. The key observation should be to determine whether the patient can rest and sleep and still maintain an adequate airway.

Feeding Problems. Because of the abnormal position of the mandible and tongue and the presence of respiratory obstruction and cleft palate, feeding becomes a troublesome matter. These children are difficult to feed; they eat small amounts, regurgitate readily, and aspirate frequently. Failure to gain weight, malnutrition, and repeated respiratory infection may be the only signs since respiratory obstruction is not always present.

Other Types of Tongue Displacement. It is not unusual in severe cases associated with cleft palate to see the tongue displaced into the nasopharynx through the cleft in the soft palate, where it is likely to cause considerable airway obstruction. Routledge (1960) has
pointed out that when the tongue is in this position, not only is greater obstruction produced, but also the obstruction is likely to increase in severity. In the neonatal period the tongue grows rapidly, and in this abnormal position growth causes the obstruction to become more severe. Even though an infant has no symptoms at the time of birth, the obstruction may become quite severe by the age of 7 or 8 weeks.

The size and position of the tongue vary greatly. Many babies appear to have a microglossus (small tongue). Others because of the relatively small jaw are described as having a macroglossus (large tongue). In time, both the large and small tongue apparently approach normal size; this disproportion has not been reported with any regularity in older children. Tongue-tie has been described with micrognathia and even with airway obstruction (Fraser and Calnan, 1961; Bromberg and associates, 1961). When the tongue and the mandible are in extreme retroposition, the floor of the mouth frequently bulges within the mandibular arch, and the tongue appears to be covering a tumor mass. The deformity usually disappears when the tongue is retracted.

Other Causes of Respiratory Obstruction. Choanal atresia can also cause respiratory problems in the newborn and has been described in association with micrognathia (Bromberg and others, 1961). It should be looked for in all such patients, and can be ruled out by passing a small catheter through each nostril (see Chapter 29). Tracheoesophageal fistula is another cause of respiratory embarrassment in the newborn, and if in doubt, should be ruled out by appropriate examinations.

Associated Congenital Defects. A number of other malformations have been reported in patients with micrognathia. In 26 cases of micrognathia, Smith and Stowe (1961) reported nine patients with cardiac murmurs, five of whom died of congenital heart disease; eight patients with anomalies of fingers and toes; three with major ear defects; two with microcephaly and mental retardation; five with serious mental retardation, and nine with major ocular anomalies. It was their recommendation that all patients with micrognathia should have a thorough ophthalmologic examination before the age of one year, including an ophthalmoscopic examination and tonometry under anesthesia, to uncover possible lesions that might be unsuspected, though treatable, and severe enough to cause blindness if not treated. The author has not encountered this high percentage of opthalmic disease, but Opitz, France, and Hermann (1972) have called attention to the frequency with which the Stickler syndrome is associated with the Robin anomalad. The Stickler syndrome or "hereditary progressive arthro-opthalmopathy" includes myopia in infancy, retinal detachment, preventable blindness, and cataracts in some cases. This is a mesenchymal dysplasia; it is an autosomal dominant mutation and should be looked for in these patients. Shah, Pruzansky, and Harris (1970) have also reported cor pulmonale in a significant number of these patients.

Treatment

The early treatment of micrognathia falls into several categories, depending on the severity of the respiratory difficulties and the feeding problems. No treatment is required if the airway obstruction and feeding difficulties are minimal. In more severe cases, if the baby is placed in the prone position, the tongue and jaw will fall forward, and this may be sufficient to overcome a slight respiratory obstruction.
As mentioned previously, the best criterion for an adequate airway is to determine whether or not the baby can breathe without obstruction while resting. If the child must struggle, cry, and stay awake in order to breathe, he will die of exhaustion unless treated.

If a change to the face down position does not suffice, the next step is to fix the tongue in an anterior position. A traction suture may be used as a temporary measure; a towel clip accomplishes the same objective and is somewhat easier to apply. In any case, a deep bite should be taken well back from the tip. A large safety pin can even be used in an emergency. A soft tube can also be inserted into the nose as a nasopharyngeal airway with good results, though it is likely to become obstructed with secretions very quickly.

If a traction suture or towel clip is needed, the patient usually will require later surgery of a more permanent nature to hold the tongue forward, since a traction suture or towel clip usually tears through tongue tissue within a period of few days. This can be done in several ways. The tongue can be sutured to the inner side of the lower lip, or a Kirschner wire can be placed through the angles of the mandible, transfixing the tongue in a forward position. Several other methods of achieving the same purpose have also been described, but these two techniques appear to be the most dependable.

In recent years maintaining the child in the face down position has gained popularity. This can be achieved by taping a stockinet to the child's head in the form of a cap and suspending the head just clear of the mattress. It can also be accomplished by using a special cut-out mattress. Such treatment requires vigilant nursing care and often prolonged hospitalization, but with the development of pediatric intensive care units, this method of treatment appears to be used more frequently (Takagi, McCalla and Bosma, 1966).

An operation is almost inevitable in a patient with micrognathia if there is a history of cyanotic attacks or of repeated severe respiratory tract infections, if the mandibular arch is more than 1 cm posterior to the maxillary arch or if the patient fails to gain weight (Robin, 1934; Benavent and Ramos-Oller, 1958; Routledge, 1960; Bromberg, Pasternak, Walden and Rubin, 1961).

Occasionally the operations designed to hold the tongue forward are not sufficient, and a tracheotomy must be done. This procedure should not be taken lightly in a newborn child because it is a difficult operation at this age even under the best of conditions. After the tracheostomy tube is in place, there is always an appreciable amount of tracheal mucosal edema, and even a millimeter of edema in the tiny trachea of a baby reduces the size of the lumen so that it is usually impossible to dispense with the tracheostomy until the child is 8 to 10 months of age or even older. This requires continuous hospitalization or very competent home care (Oeconomopoulos, 1960; Moyson, 1961; Stool, Campbell and Johnson, 1968).

Many procedures have been suggested which pull or push the mandible forward, but in general they have proved to be ineffective, cumbersome, and at times dangerous (Callister, 1937; Longmire and Sanford, 1949; Routledge, 1960). Simple gadgets to be attached to the feeding bottles have been designed for the purpose of making the child thrust his jaw forward. They may be partly effective, but cannot be relied upon to treat respiratory obstruction (Davis and Dunn, 1933). Robin was much concerned with the need for what he termed "orthostatic" feeding; this consisted of holding the baby in a nearly vertical position, particularly while
nursing, so that he was forced to push his jaw forward (Robin, 1927b). Gavage may be needed as a temporary measure, particularly if there is a tendency to aspirate.

Douglas popularized an operation described earlier by Shukowsky (Shukowsky, 1911; Douglas, 1946, 1953). This procedure consists of a tongue to lip suture. In reporting his own cases, in addition to information obtained by a questionnaire, Douglas noted only one death (a premature infant) in 31 patients operated upon. In 21 patients not operated upon but treated conservatively, there was a 65 per cent mortality (Douglas, 1950). Kiskadden and Dietrich (1953) reported a 30 per cent mortality in 15 patients treated without operation and none in 10 patients who were operated upon. Similar series have been reported by others (Beers and Pruzansky, 1955; Routledge, 1960; Woolf, Georgiade, and Pickrell, 1960; Bromberg and coworkers, 1961).

Another effective method of correcting glossoptosis is to use a Kirschner wire which is placed through the mandible just anterior to the angle, transfixing the tongue and passing on through the mandible on the opposite side. Hadley (1961) reported using this in a child whose tongue had been badly torn.

Routledge (1960) pointed out that in the Douglas procedure (in which the under surface of the tongue, the central portion of the floor of the mouth, the alveolar ridge, the buccal sulcus, and the labial mucosa are denuded to allow the adherence of the tongue and its maintenance in a forward position), the degree of fixation is small, the submaxillary duct openings are jeopardized, and it is difficult to obtain a satisfactory adhesion. Routledge has proposed a different method of tongue-lip fixation which has been further modified and is described in detail later in the text.

McEvitt (1968, 1973) reported success by passing a nasogastric tube. This tends to hold the tongue a little forward; it also allows feeding by gavage, but most importantly it apparently provides a significant airway on either side (laterally) of the tube. Our experience with this technique is small and includes one or two failures.

Burston (1966) favored a prolonged hospitalization with a frame to hold the child in the face down position. This, as noted above, has a measure of success, but several months' hospitalization is not practical in many areas.

Initially Robin described a "monobloc" to obturate the cleft and to hold the tongue and mandible forward. From time to time the variations on this approach have been described. Fára (1974) found this technique sufficient in about two-thirds of his patients and emphasized the advantages of preventing the tongue from becoming displaced into the cleft where it obturates the airway.

**Operative Technique.** General anesthesia, preferably with an endotracheal tube in place, is desirable if the services of an expert anesthesiologist are available. As has been pointed out, tracheal intubation in a baby with retrognathia is an extremely difficult procedure and is preferably done with the infant awake, since complete obstruction may occur as soon as relaxation is produced by the anesthesia. If tracheal intubation is not successful, anesthesia can be maintained by insufflation with a nasopharyngeal tube plus the careful use of a tongue suture to maintain an adequate airway.
The basis of the procedure described by Routledge is a horizontal attachment rather than a longitudinal one. When the tongue is placed forward against the lower lip, contact is established between a broad expanse of the lower lip and the free edge of the tongue. These two surfaces are incised (after injecting a weak solution of epinephrine for hemostasis). The inferior edges of the incised structures are approximated with a running 5-0 chromic catgut suture evertting the mucosal edges.

A deep suture of 0 or 00 silk is next placed below the chin pad, brought up anteriorly to the mandibular symphysis and out through the lip incision. A suture carrier or Reverdin needle is used to place it through the tongue incision and back through the substance of the tongue as far posteriorly as possible. The suture is then threaded through a medium sized button, brought back through the tongue and lip and through a button beneath the chin. A retrieving suture is also placed through the tongue button, brought out through the mouth, and taped to the cheek.

Two sutures of 3-0 chromic catgut approximate the muscles of the tongue and lip, one on either side of the silk suture, and the superior mucosal edges are approximated with a continuous 5-0 chromic suture evertting the edges. Placing the chin button below the chin pad minimizes scarring without changing its effectiveness. The button suture is left in place for 7 to 10 days and the lip adhesion maintained for 10 to 18 months. When a tongue-tie is present, it must be relieved before the tongue can be brought forward. Gavage is done for feeding for the first week postoperatively to minimize tongue movement (Randall and Hamilton, 1971).

Other techniques have been described by Blocker and Lewis (1954), who employed fascia lata as a deep suture through the base of the tongue, holding it forward to the symphysis. To transfix the tongue accurately with a Kirschner wire, Schatten and Tidmore (1966) used a tongue suture and withdrew the endotracheal tube in the nasopharynx. (The same technique can be used with a nasopharyngeal insufflation.) The Kirschner wire is inserted through the skin and the mandible superior and anterior to the angle but not so far as to injure the inferior alveolar nerve. The tongue is retracted from the mouth to a position just beyond where it causes obstruction to the airway. The Kirschner wire is then placed through the tongue and into the mandible on the opposite side. The tongue suture can then be relaxed to see if a satisfactory airway can be maintained. If the tongue is pulled too far anteriorly at the time it is transfixed, swallowing becomes very difficult. The end of the Kirschner wire is left protruding from the skin and protected by adhesive tape or is cut just short enough to be buried by the skin but long enough so that a small incision can later be made to permit its withdrawal.

Oeconomopoulos (1960) reported success with a suture placed horizontally through the base of the tongue and brought forward, transfixing the alveolus.

Wang and Macomber (1963) described a variation of the Douglas procedure. They raised flaps of mucosa, one from the under surface of the tongue, the other from the buccal surface of the lip. These flaps are interdigitated, thereby giving a broader surface of contact than is achieved by the Douglas procedure, and, at the same time, avoiding the openings of the submaxillary ducts and the need to remove alveolar mucosa.
Growth and Development

Robin felt that the respiratory insufficiency caused by glossoptosis would lead to physical backwardness and mental retardation, and to a child easily angered, difficult to control, and unable to concentrate, with headache, nightmares, and other disturbances which would persist through infancy, childhood, and adult life (Robin, 1926c, 1934). These complications probably occur only in the patient with brain damage.

The growth potential of the micrognathic jaw of the Pierre Robin anomalad is inconsistent. A number of authors have felt that the mandible will reach normal proportions in all cases, but a definitely diminished growth potential can be demonstrated in some (Randall, Krogman and Jahina, 1964). Patients with similar retrognathic positions may develop along one of four growth patterns: (1) normal growth of mandible and maxilla with eventual normal dental occlusion; (2) diminished mandibular growth with normal maxillary growth but with forward protrusion of the mandible to produce fairly satisfactory anterior dental relationship; (3) diminished mandibular growth, normally maxillary growth, and a severe Class II malocclusion; (4) diminished mandibular growth and diminished maxillary growth with slight Class II malocclusion (Chapple, 1935; Sjolin, 1950; Pruzansky and Richmond, 1954; Bromberg and coworkers, 1961; Krogman, 1962; Randall, Krogman and Jahina, 1964).

The cleft palate, when present, almost always involves only the soft palate or the soft palate and the posterior portion of the hard palate, and can be closed in the usual manner. In addition to the obvious cleft, there usually appears to be a severe amount of shortening of the anteroposterior dimension of the palate. Orthodontics, cartilage implants to the chin, and repositioning of the mandible by osteotomy may each have a place in the late correction of the micrognathia.

Complications

Possible complications fall into two categories. There are those which are caused by the respiratory obstruction when it is not treated adequately, and those which are the result of the suggested methods of treatment.

In severe cases of the Robin anomalad, death from respiratory obstruction can occur immediately if the situation is not recognized and the problem corrected. More likely the respiratory obstruction will be partial and can be overcome by the child when he cries and strains. As noted earlier, if the child's airway becomes obstructed when he relaxes, it will be necessary for him to strain in order to breathe, and under these conditions he can literally exhaust himself to death. The down hill course can be quite insidious and the termination very abrupt. With lesser degrees of obstruction, the complications can include aspiration with episodes of pulmonary infection and poor eating with failure to gain weight. Often, the respiratory obstruction is intermittent and leads to episodes of cyanosis. Procrastination in alleviating this situation can lead to accumulating increments of cerebral anoxia, resulting in a child with permanent brain damage.

The innumerable possible complications alluded to by Robin are noted under Historical Aspects. It is surprising in recent experience that the problem of respiratory obstruction does
not always improve as the child grows. During the first three months of life improvement is usually seen, but the situation can also become steadily worse. This is most often seen in the child whose airway obstruction is not his most serious problem but whose poor nutrition and respiratory infection are the salient features. Occasionally a child is seen in whom a disproportionate growth of the tongue and the nasopharynx occurs, with more growth in the former than in the latter. Under these conditions the degree of respiratory obstruction can gradually increase.

The face down position can lead to chronic skin breakdown if this is not watched for carefully. The suspensory headcap has also been a problem in that occasionally it will constrict the bones of the calvarium and cause overlapping of their edges.

Complications of the surgical techniques have been alluded to, but it should be restated that if a traction suture is used in the tongue for more than a day or two, it is very likely to cut through and leave the tongue torn and infected. Should this occur, it is probably best to maintain the airway either with the head down position or with the Kirschner wire transfixion of the base of the tongue. Immediate suture of the lacerated tongue is achieved only if the wound is fairly clean. This is not usual. Ordinarily it is better to let the tissue to heal and to perform a secondary repair a year or two later.

Should the tongue-to-lip adhesion break down, it is unlikely that a second attempt of surgery in this area will succeed, since these wounds become quickly reinfected. As a result, one of the other methods of treatment noted above should be employed. In the use of the Kirschner wire through the angles of the mandible, skin breakdown over the wire is a frequent complication and can lead to appreciable scarring in the skin of the cheek. To avoid this the ends of the wires can be left buried, but it is important to keep the baby from lying on the side of his cheek, a position which would produce pressure directly over the end of the pins.

It bears repeating that sudden death following exhaustion is a frequent complication of inadequately treated Pierre Robin anomalad. Also, repeated episodes of partial obstruction with cyanosis can lead to permanent brain damage (Hoffman, Kahn and Seitchik, 1965; Randall and Hamilton, 1971).