UNILATERAL DIAPHRAGMATIC PARALYSIS AND ERB'S PALSY IN THE NEWBORN

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Since Naunyn reported the first case of unilateral diaphragmatic paralysis in a newborn infant in 1902, 38 further cases have been described; 72% showed an association with Erb's palsy on the same side as the phrenic injury.

As most cases have occurred after breech delivery, the majority of observers considered the injury to result from manipulations of the head and neck producing overstretching of the cervical nerves. Rupilius (1934), however, believed that malposition of the foetus in utero allowed compression of the brachial plexus and phrenic nerve between the clavicle, the first rib, and the transverse processes of the fifth and sixth cervical vertebrae.

The present case showed avulsion of the anterior roots of the cervical nerves supplying the phrenic nerve on the affected side indicating that trauma by traction had occurred at birth.

Case History

L.T., a boy, aged 47 days, was born at term on January 7, 1952, of a primiparous mother after a difficult breech delivery during which forceps were applied to the after-coming head. A right-sided Erb's palsy was noted soon after birth. His condition was poor for the first two days of life but then became satisfactory. He was not weighed at birth but his weight at 3 days was 7 lb. 5½ oz. (3,325 g.). When 9 days old periods of rapid breathing were noticed, feeds were taken slowly and he tired easily. He was discharged from the maternity department when 15 days old, feeding from the breast.

Physical examination when the infant was aged 16 days (weight 7 lb. 5½ oz., 3,325 g.) showed that he was vigorous with a good colour and a typical right Erb's palsy. Respirations varied between 50 and 70 per minute, and while watched in the Out-Patient Department of the Queen Elizabeth Hospital there were periods of rapid breathing separated by periods of slower, quieter breathing. There was no rib recession, the percussion note was everywhere resonant, and the breath sounds were normal. The liver was of normal size, the spleen was just palpable and all other systems were normal.

The mother was feeding her infant two-hourly chiefly with the bottle and the baby had apparently taken only about 13 or 14 ounces in 24 hours. He was admitted to hospital on account of his rapid respiratory rate.

A radiograph of the thorax showed collapse of the right lower lobe with a possibly raised right dome of the diaphragm or an encysted effusion, and screening showed paradoxical movement of the right diaphragm (Dr. C. Hodson). A diagnosis of right diaphragmatic paralysis was made. When the baby was 24 days old the respiratory rate still varied considerably and intermittently. The upper part of the right thorax moved more and the lower part less than the left chest. There was a little subcostal recession on the right and the percussion note was impaired and air entry diminished at the right base posteriorly. The child's condition gradually deteriorated with increasing dullness at the right base and a persistent increase of the respiratory rate. The amplitude of the right thoracic movements increased with the lower part swinging upwards and outwards on inspiration, accompanied by indrawing of the epigastrum.

Cyanotic attacks occurred on the twenty-sixth and twenty-seventh days of life and on the following day the child had generalized twitching. A radiograph of the thorax showed a slight increase in the height of the right dome of the diaphragm. The blood showed a leucocytosis of 22,000 per c.mm. (polymorphs 33%, lymphocytes 63%, monocytes 4%). His colour and vigour continued to deteriorate, his breathing was distressed, and oxygen therapy was given. When 31 days old, tube feeding was necessary and was continued thereafter. At 36 days he was dyspnoeic and cyanosed after exertion and showed bilateral rib recession, a grossly impaired percussion note at the right base and mid-zone with absent breath sounds at the right base and numerous crepitations in the right mid-zone. His leucocyte count was 26,000 per c.mm. (polymorphs 37%, eosinophils 1%, lymphocytes 52%, monocytes 10%). Adventitious sounds increased and by the forty-second day crepitations were also heard at the left axilla and base. On the forty-seventh day artificial respiration in a Drinker respirator was given; he rapidly deteriorated and died 12 hours later.

He received oral penicillin from the seventeenth day
of life, chloramphenicol from the thirty-third day and intramuscular streptomycin from the thirty-sixth day; none had any appreciable effect on his condition.

Necropsy Findings. The body was that of a white male infant weighing 8 lb. 4½ oz. (3,750 g.) and measuring 55 cm. in length. There was marked flattening of the right side of the thorax and the abdomen was scaphoid.

Both lungs (R. 27 g., L. 36 g.) occupied the posterior halves of their respective pleural sacs and showed extensive areas of collapse posteriorly. This was noticeable on the right side where the dome of the diaphragm extended to the level of the first intercostal space.

The heart (35 g.) extended for 4.5 cm. to the left of the midline and 1.0 cm. to the right and was normal in all respects.

The adrenals together weighed 7.4 g. and were distinctly thicker than normal. On the cut surface neither showed any obvious zonal arrangement and the entire section was uniformly pale yellow, opaque and friable.

Dissection of the neck showed a normal brachial plexus on the left side. On the right, however, there was complete avulsion of the anterior nerve roots of the third, fourth and fifth right cervical nerves at their attachment to the spinal cord (Fig. 1). Opposite these nerves the epidural space was widened and the dura itself thickened although there was no evidence of old or recent haemorrhage. The right phrenic nerve, which arose mainly from the third cervical nerve and received a twig from the fourth cervical nerve, was intact throughout its course.

There was a small recent haemorrhage in the region of the lateral cord of the right brachial plexus but the plexus itself was uninjured.

All other organs were grossly normal.

Microscopical Examination. Degeneration was demonstrated in the right fourth cervical, phrenic and musculo-cutaneous nerves, using the left fourth cervical nerve as a control.

The muscle of the right dome of the diaphragm was very thin and the individual fibres small and atrophic, giving a rather variable reaction to basophilic stains.

Sections from both lungs showed extensive collapse only.

Both adrenal glands showed extensive necrosis throughout all zones (Fig. 2). The central area resembled necrotic foetal cortex but the process also involved much of the definitive cortex even extending to the capsule over large areas. Where the definitive cortex was intact, glomerular and fascicular zones could be distinguished, often separated from the necrotic region by condensed connective tissue containing numerous haemosiderin-laden phagocytes. Throughout the necrotic areas irregular foci of calcification of varying size were scattered. No thrombosed vessels were demonstrated.

Discussion

Of 29 cases in which diaphragmatic paralysis was associated with Erb's palsy, including the present
case, eight (27.6%) died; only one death (9%) occurred in 11 infants in whom there was no accompanying brachial palsy (Blattner, 1942). Necropsy findings were available in only six cases in the literature. All patients died between 2 and 6 months of age and the lesion was right-sided in all cases. Elevation of the right dome of the diaphragm with extensive collapse of the right lung was always present. The brachial plexus was dissected in three infants (Landsberger, 1926; Rupilius, 1934; Turner and Bakst, 1949) but showed no abnormality. The phrenic nerve showed evidence of degeneration in three cases (Cocchi, 1937; Blattner, 1942; Turner and Bakst, 1949) but was not examined microscopically in the rest. In Tyson and Bowman's case (1933) death was attributed to congenital heart disease and the brachial plexus was not explored.

The only case in which the cervical nerve roots were examined was that of Turner and Bakst (1949) which was also reported by London (1949) and Schifrin (1952). This was a 13-week-old male who had been born by breech presentation with the assistance of forceps. A right-sided Erb's palsy was noted soon after birth; respiratory difficulty, at first intermittent, as in the present case, became continuous and more severe and at the age of 6 weeks diaphragmatic paralysis was diagnosed. At necropsy the brachial plexus was normal but when the nerve roots were traced into the spinal canal avulsion of the anterior roots of the fifth and sixth cervical nerves at their junctions with the spinal cord was revealed. The phrenic nerve, which arose mainly from the fifth cervical nerve with a small twig from the fourth cervical nerve, was completely demyelinated.

Consideration of the similar findings in Turner and Bakst's case (1949) and in the present case suggests the probability that when diaphragmatic paralysis and brachial palsy co-exist the site of injury is the anterior roots of the cervical nerves and, as this lies within the membranes covering the spinal cord, it is presumably caused by tension rather than by pressure. When complete avulsion has occurred regeneration of the nerves is probably impossible. Since the majority of cases improve or recover complete movements it is likely that minor trauma may result in rupture of nerve fibres without wide separation of their torn ends.

The other interesting feature of the present case was the incidental finding at necropsy of extensive destruction and calcification of both adrenal glands. Although tuberculous foci in the adrenals with subsequent Addison's disease are probably the commonest cause of calcification, this is extremely rare in childhood (Chenoweth, 1948). Neuroblastoma (Willis, 1948), phaeochromocytoma (Bartles and Cattell, 1950) or adenocarcinoma of the cortex (Samuel, 1948) may show calcium deposits in their substance. It is probable, however, that adrenal calcification in infants and young children is usually a sequel to sublethal haemorrhage occurring in the neonatal period. Calcification and haemorrhage have been found together in a child as young as 8 days while in older children previous haemorrhage is suggested by the presence of haemosiderophages in the region of the calcified areas (Snelling and Erb, 1935).

Summary

Intermittent rapid breathing in a 16-day-old infant with Erb's palsy led to the radiological diagnosis of right diaphragmatic paralysis. During the remaining 31 days of life respiratory distress increased and evidence of pulmonary collapse appeared.

At necropsy avulsion of the anterior roots of the cervical nerves supplying the right phrenic nerve was found. Adrenal calcification following old haemorrhage was also present.

Previously described necropsy records are reviewed.

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References