Kinking of the medulla in children with acute cerebral oedema and hydrocephalus and its relationship to the dentate ligaments

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While developing a method for doing perinatal necropsies some 4,000 necropsies ago, we were disturbed about the contamination of the cerebrospinal fluid with blood whilst opening the skull, and a technique was devised of opening the dura over the base of the skull, sampling the cerebrospinal fluid and inspecting the foramen magnum from below before the skull was touched.

That method (Emery, 1960) has been applied as a routine procedure at the Sheffield Children's Hospital for over 10 years. It enabled us to find the severe brain swelling that sometimes occurs in children following relatively minor burns (Emery and Reid, 1962). The technique has also brought to our notice instances in which death was apparently associated with kinking of the brain-stem at its junction with the spinal cord, a lesion that we have not found described previously in presumably healthy children.

This bending of the brain-stem appears to be due to a downward and backward displacement of the intracerebral brain-stem while the spinal cord is held rigid by the dentate ligaments. This deformity is seen as an acute lesion in children dying in a variety of diseases associated with unconsciousness and convulsions. It also has close analogies with the spur and 'Knickung' deformity forming part of the 'Cleland' or 'Arnold-Chiari' malformation (Cleland, 1883; Schwalbe and Gredig, 1906). It seems likely that the post-natal acute lesions may illustrate the mechanism by which the spur part of the Chiari type II deformity develops in utero.

In this paper, illustrative cases only are described for which photographs are available. Only two cases of Arnold-Chiari deformity are illustrated from a series of over 200 brains of this type that we have examined.

CASE 1 (N.4481) This child died at the age of 3 years. She had been a completely normal child until six days before death when she complained of a pain in the left leg. She was off colour and the following day began to vomit. She commenced convulsing and was admitted to hospital with a temperature of 103°F. and a diagnosis of febrile convulsions.

She remained very drowsy and lumbar puncture revealed clear, normal fluid. The swelling of the left leg increased and the possibility of osteomyelitis was considered. She was then transferred to the Children's Hospital where she was found to be unconscious but responded to painful stimuli. A diagnosis of osteomyelitis with possible cerebral abscesses was made.

The child was taken to the theatre. As she was already unconscious, no anaesthetic was required and a subperiosteal abscess of the left tibia was opened. The most remarkable thing about her clinical state was the variable level of consciousness. Extensive biochemical tests revealed no significant changes. She died three days later.

Necropsy showed an extensive osteomyelitis of the tibia but no abscesses or sepsis elsewhere.

FIG. 1. The exposed brain-stem below the foramen magnum. The poles of the cerebellum extend well below the skull and one of the posterior inferior cerebellar vessels can be seen on the near side. The caudal displacement of the dorsal cervical roots can be seen below this vessel. (Photograph from a colour transparency taken during necropsy.)
Opening the dura beneath the foramen magnum revealed virtually no free fluid. The brain-stem was impinging on the posterior surface of the spinal theca. Two leaves of cerebellum were protruding through the foramen magnum (Fig. 1) and the brain-stem appeared to begin as a round mass protruding below the cerebellum.

When the spinal cord was cut in the mid-cervical region, there was no gaping but, when the dentate ligaments were removed, the knuckle of the medulla diminished and the upper cervical cord moved down, overlapping the remaining cord.

When the skull was opened the brain was found to be symmetrical with gross general flattening of the convolutions and no free cerebrospinal fluid. The lateral ventricles were reduced in size. There was a small amount of herniation of the cerebral hemispheres against the brain-stem and down through the tentorium. No cerebral abscesses or local areas of softening could be found.

When the brain-stem was removed from the skull with the cerebellum, the kink in the brain-stem was not obvious, and if it had not been observed previously when the dura over the fourth ventricle was opened before opening the skull, the lesion could very well have been missed. When, however, the spinal cord was held and gently pushed towards the brain, the deformity was immediately reproduced. A lateral view of the deformity is shown in Figure 2.

The unconsciousness and the probable cause of death in this child appears to have been due to a severe compression of the medulla. The kinking of the brain-stem was undoubtedly due to its caudal displacement. That the dentate ligaments had restricted the movement of the brain-stem showed clearly when they were cut. The kinking of the brain-stem would not have been found if the skull had been opened and the brain removed in the conventional way.

CASE 2 (N.3917) This child, who died at the age of 6 weeks, was born at full term and apparently was completely normal until four days before death when she vomited, had her bowels open twice, and had a possible minor convulsion.

She was admitted to hospital with a temperature of 103°F. There was hyperextension of the back and the left arm was twitching. The cerebrospinal fluid was clear. There was no increase in white cells or alteration in sugar but there were 240 R.B.C.s per c.mm. and the protein level was 200 mg./ml. At that time, the anterior fontanelle was reported to be tense by one physician and normal by another.

She was given large doses of antibiotics and treated as a case of meningitis. The next day her condition became worse and the cerebrospinal fluid showed the presence of 8,000 nucleated cells per c.mm., most of which were polymorphonuclears, but no organisms could be seen and the sugar content was normal.

Her general condition deteriorated, the limbs gradually becoming more and more spastic until death occurred a day later.

Necropsy revealed no gross abnormalities outside the skull and central nervous system.

Opening the dura at the back of the neck revealed a pale mass with very little free cerebrospinal fluid and that which was present was slightly bloodstained.

Within the skull there was antemortem thrombosis of the longitudinal sinus and recent thrombosis of both lateral sinuses; the thrombosis of the former was obviously the older. The straight sinus also appeared to be the site of recent thrombosis. The surface of the cerebral hemispheres was pale and there was gross flattening. The whole of the centre of the brain was the site of haemorrhagic disorganization (Fig. 3).
The thrombosis and haemorrhage had been associated with a gross increase in bulk of the brain, forcing the brain and brain-stem down into the neck, thus producing a knuckle visible through the dura at the back of the neck at necropsy. It is possible that this knuckle accounted for the child’s progressive spasticity.

CASE 3 (N.4247) This child, a girl, aged 3 years 4 months, had sustained a road accident and the wheel of a car went over her abdomen. She was admitted unconscious, requiring an immediate tracheostomy, and died three hours later, six hours after the accident.

At necropsy the main evidence of direct trauma was found to be in the abdomen. She had a laceration of the liver, haemoperitoneum, haemorrhage in the right adrenal, fracture of the clavicle, and there were multiple haemorrhages in the right lung. When the base of the skull was opened at the back of the neck, a knuckle of the lower medulla was seen pressing against the dura and the cerebellum was just visible in the foramen magnum (Fig. 5). A small hypodermic needle was stuck straight into the cord, the direction of the needle being, as far as could be judged, parallel to the plane of the foramen magnum.

When the cerebellum and brain-stem had been removed, it was seen that the pin had prevented the complete spontaneous straightening of the brain-stem and the head of the hypodermic needle was pressing quite firmly against the cerebellum (Fig. 6).

The brain was removed with a length of spinal cord. Lying in the saline the cord appeared straight, but when it was gently approximated to the brain-stem, an acute knuckle occurred just below the cerebellum and exactly reproducing the situation seen when the dura over the base of the skull had been opened during necropsy (Figs. 4a and b).

This child had died as a result of a saggital sinus thrombosis with a terminal thrombosis of the central cerebral veins.

The manoeuvre with the hypodermic needle illustrates to some extent the way in which the kinking present in a cadaver undoes itself when the brain
No free cerebrospinal fluid was visible through the posterior incision.

Opening the skull bones revealed no free fluid but an extremely tense brain with obvious gross swelling of the cerebral hemispheres. The inferior lips of the temporal lobes were compressing the peduncles and a short distance through the tentorium.

The cerebellum appeared to be compressed but did not enter the foramen magnum. The spinal cord in the region of the medulla showed an acute backward kink. The kinking in this case was different from those in the

FIG. 6. Photograph of the cerebellum and brain-stem of case 5.

The needle had been inserted at necropsy parallel to the foramen magnum. At that time it was well clear of the cerebellum. The needle came to press against the cerebellum after removal from the skull.

The photograph shows how the natural straightening of the kinked brain-stem had been restricted by the needle.

and brain-stem are removed from the skull and spinal column.

The brain in this child showed no evidence of haemorrhage or bruising but simply severe flattening of the convolutions associated with general swelling.

CASE 4 (N.4159) This was a full-term child weighing 5 lb. 10 oz. At birth there had been difficulty with breathing and she had never been very lively.

She was admitted one day before death with a temperature of 95°F. with rales in the chest and occasional bouts of coughing. She was nursed in an incubator but had repeated convulsions with clonic movements of the arms and legs. There was no hyperglycaemia. A clinical diagnosis was made of a salt-losing syndrome associated with water intoxication. The child died within a few hours of admission, three weeks after birth.

Necropsy showed a normally formed child with oedema of the buttocks and back. The heart was grossly enlarged and she had obviously had chronic cardiac failure associated with a ventricular septal defect. The immediate cause of death appeared to have been due to extensive haemorrhages in the lung, and histology confirmed the presence of an acute pneumonia.

Opening the cervical spine at the back of the neck revealed the presence of a pale mass coming directly down from the posterior lip of the foramen magnum. This mass extended into the neck for a distance of 9 mm.

FIG. 7a and b. Posterior (a) and lateral (b) views of the junction of the brain-stem with the medulla in case 4.

The deformity did not undo itself when the tissues were removed from the skull. Note also the general compression of the cerebellum.
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previous cases in that it did not disappear spontaneously and Figs. 7a and 7b show the posterior and lateral view of the brain-stem with the specimen fixed in a free state. The cerebral hemispheres showed no gross haemorrhages or tumour and there was no hydrocephalus, simply generalized swelling.

This case differs from the previous cases in that the child had undoubtedly been ill since birth and had had cerebral hypoxia at that time. The possibility that the lesion may have been present before birth and may have accounted for the child's difficulty in breathing and later death cannot be excluded.

CASE 5 (N.4441) This girl had chronic spastic diplegia. The condition followed an incident in infancy which was thought to have been a thrombosis of the longitudinal sinus.

The child died at the age of 9 years, four days after she had been admitted to hospital for operative dental treatment which had involved a short general anaesthetic. Although she was off her food, she was not pyrexial and she was discharged home.

She had a cough but no particular notice was taken of this. Two days later as her 'chestiness' was increasing, she was admitted to hospital, cyanosed, partially unconscious but apyrexial. Sucking out mucus from the larynx and trachea appeared to help her temporarily but within a few hours she commenced to have phasic respirations and became fully unconscious. Her condition steadily deteriorated to death. Clinically, no explanation for her condition was forthcoming.

At necropsy, the only lesion found in the viscera was a small amount of fatty change in the liver; there was no pneumonia. The most striking feature, however, was the presence of a marked kinking in the brain-stem associated with symmetrical partial herniation of the cerebellar tonsils. The posterior wall of the medulla in the region of the kink was displaced backwards beneath the foramen magnum so that it was in contact with the dura at the posterior wall.

The brain showed a mild degree of hydrocephalus affecting chiefly the lateral ventricles. The aqueduct of Sylvius was patent. There was no evidence of any narrowing or obstruction of the longitudinal sinus.

Figure 8 shows a longitudinal cut through the cerebellum and medulla compared with a similar cut from a child's brain of approximately the same age. Obvious abnormalities are the approximation of the cerebellum to the posterior surface of the medulla apparently associated with a shifting back and compression of the medulla in the region of the inferior olives.

The precise mechanism of death in this child was obscure, even after full necropsy. There were some microscopic haemorrhages in the cerebral peduncles but these were simply of terminal origin and there was no longstanding compression of the cerebral peduncles. The kinking of the brain-stem at the lower end of the fourth ventricle did not spontaneously resolve and thus had probably been present for a considerable time before the child's death. An explanation for this child's death is that there was fluid retention within the brain and ventricles associated with an acute phase of intracranial hypertension with further compression of the lower medulla.

CASE 6 (N.3418) This child died at the age of 10 days with peritonitis and a ruptured bladder. She was of 36 weeks' maturity, weighed 6 lb., and was admitted to hospital on the day of her birth because of a thoraco-lumbar meningomyelocele and congenital hydrocephalus. There was paraplegia and incontinence.

The meningomyelocele was repaired on the day of birth, but a week after operation she commenced vomiting bile-stained fluid, and the abdomen became distended. The blood urea level was 102 mg.%. Her general condition steadily deteriorated to death which was due to a rupture of a diverticulum of the bladder with terminal peritonitis.

Dissection of the back of the neck showed a spur descending for a distance of 44 mm. beyond the foramen magnum and the fourth ventricle extended for a distance...
of 22 mm. overriding posteriorly. The cerebellum came down below the foramen magnum for a distance of 10 mm. (Fig. 9).

CASE 7 (N.3291) This child was born with a meningo-myelocoele 'plaque' in the region of L3-S1 with an abnormal cauda equina and coccyx. He died at the age of 8 weeks with a purulent ventriculitis associated with a pyocyaneus infection. The appearance of the dissected cervical spine is shown in Figure 10.

The 'spur' extended 28 mm. from the skull, overriding the cervical cord for a distance of 5 mm. The cerebellum

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**FIG. 9.** Photograph of a dissected cervical spine from a child (case 6) with meningo-myelocoele and hydrocephalus in which the upper dorsal roots have been removed and the dentate ligament on the right side has been exposed. The tip of the medullary spur has been opened revealing the communication with the fourth ventricle. The posterior aspect of the spur has been similarly opened, revealing the mass of choroid plexus and connective tissue overlying this area. The atlanto-occipital joint (A) is exposed on the right side of the photograph just beside the dorsal root ganglion of the first cervical nerve.

Note the thick attachment of the dentate ligament to the skull just within the foramen magnum. Note the way in which the attachments of the dentate ligament communicate over the back of the cord to form a sling-like structure (see arrow). This restrains the brain-stem and the medullary spur over-rides it posteriorly.

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**FIG. 10.** Photograph of the exposed cord from a child (case 7) with hydrocephalus and meningo-myelocoele in which the dorsal roots have been removed from the upper cervical segment and the dentate ligament has been exposed.

The photograph illustrates the way in which the medullary spur lips backward over the dentate ligament, the latter forming a virtual sling holding the cord taut at this point. The lateral attachments of the dentate ligaments also shows the slight downward displacement of the cord throughout the whole region photographed.
extended only a small proportion of the distance of the fourth ventricle and spur, the whole being surrounded by a thin network of fibrous strands. The relationship of the dentate ligament to the spur is obvious in Fig. 10 as also is the increased thickness of the first part of the ligament.

**DISCUSSION**

The cases presented here fall into three groups: the first contains three children in which kinking of the brain-stem occurred associated with a rapid swelling of the brain, related to an acute infection (case 1); an infection with cerebral venous thrombosis (case 2); and a road accident (case 3). In these children the kinking of the brain-stem appeared to be of very recent origin and entirely 'recoverable' in that when the brain and upper cervical spine were removed from the body the bending of the brain-stem undid itself completely.

The second group comprises two children who had more longstanding cerebral lesions. One, a child of 3 weeks who possibly had cerebral swelling at the time of birth (case 4), and the other with an almost identical brain-stem lesion, had spastic diplegia (case 5). The spinal deformities had a form similar to that in the acute cases.

In the third group are presented two children with meningomyelocele, hydrocephalus, and an Arnold-Chiari deformity associated with medullary 'spurs'.

A common feature in all cases presented here is that there appears to have been an increase in bulk of the cerebral hemispheres, producing a downward (caudal) movement of the brain-stem. Movements of the brain-stem have been recognized for many years. Johnson and Yates (1956) showed that it could move sufficiently to produce a kinking and obstruction of the posterior cerebral vessels and Dott and Blackwood (1951) had apparently made similar observations. Howell (1959 and 1961) described a series of cases in which there was compression of the upper brain-stem with foramenal impaction associated with a variety of symptoms, including decerebrate rigidity.

The brain-stem within the skull is a relatively mobile structure with no direct fibrous attachments or suspensory ligaments to the lateral walls of the skull. The spinal cord, on the other hand, is firmly suspended (Kahn, 1947) within its dural sheath by the dentate ligaments, having lateral attachments at each vertebral level and an upper attachment to the dura forming the rim of the foramen magnum. Thus, the spinal cord is held in a relatively rigid state while the brain-stem above this level is free.

Our own observations at necropsy suggest that the brain-stem is normally under a slight degree of stretch. Since we have been using the technique of opening and examining the foramen magnum before opening the skull, it has been our practice to make a transverse cut across the spinal cord immediately below the foramen magnum. When this is done in children, the two cut surfaces spring apart for a variable distance of up to 5 mm. and it is only with some difficulty that the head and neck can be manipulated to bring the two cut ends into direct apposition. The amount of movement seems to be about equal up and down but the caudal movement of the cord is no more than 2 mm.

Howell (1959) states that in an adult the dentate ligaments hold the spinal cord to allow a maximal movement of 1 cm. in the upper cervical cord. We have attempted to make measurements in this way in infants but the softness of the brain tissue makes this a little difficult. If the brain-stem is cut across above the level of the dentate ligaments the brain can usually be moved cranially for over a centimetre. If the brain-stem is cut at the level of the third or fourth cervical segment, the movement upwards cranially of the brain-stem is limited, and in a child under 2 years, a distance of about 5 mm. appears to be the maximum.

The caudal movement of the brain-stem appears to be more limited than that of cranial movement. While the dentate ligaments are attached, a downward movement of the cervical cord of more than 3 mm. is not possible without actual damage to the brain. If, however, the dentate ligaments are divided, the upper cervical cord can descend, its level of descent being limited by the cerebellum. The latter appears to interfere with the descent at a distance of about 10 mm.

The situation then seems to be that in the normal child's brain (birth to 1 year), the brain-stem is free to move in a caudal direction for a distance of about 3 mm. before it is held by the dentate ligaments.

The deformity of the brain-stem associated with the Arnold-Chiari deformity is not that of simple caudal displacement. The elongation of the fourth ventricle was described by Cleland in 1883 and Chiari in 1891, and excellent illustrations of this deformity are presented by Russell and Donald (1935) and Daniel and Strich (1958). Many theories are discussed in an extensive article by Barry, Patten, and Stewart (1957), who produce evidence against the idea that the medullary defect is due to traction on the spinal cord and suggest that the deformity is due to irregular overgrowth of the central nervous system. They consider that the 'knuckled' appearance of the medulla is due to an 'overgrowth' in length and circumference of the embryonic medulla and to its being retained by its dural attachments within the cervical canal. The literature on the causes of this deformity is considerable and has been extensively
discussed by Peach (1964), but the causes are still largely speculative.

Whatever the primary cause or causes of this deformity, there is no question that there is a gross caudal displacement of the bra'n-stem (Russell, 1949). Emery and Levick (1966) measured the relationship of the origin of the basilar artery and the base of the fourth ventricle to the skull. They showed that in eight cases of thoraco-lumbar meningo-myelocele, the vertebral artery was displaced caudally for a distance on average of 5 mm. and the base of the fourth ventricle 13 mm. In seven cases of lumbo-sacral meningo-myelocele, the average descent of the basilar artery was 10 mm. and of the fourth ventricle 21 mm. These measurements give some indication of the distortion of the medulla and the way in which the dorsal part of the medulla moves a greater distance.

The most reasonable explanation of this state is that the brain-stem is pushed down as far as it can be and when it is held, it folds over dorsally to produce the 'spur' and the 'knickung' of the German literature. The fold is backwards because of the general curvature of the cord at this level.

The restricting effect of the uppermost part of the dentate ligaments is obvious in dissected specimens as illustrated in Figures 8 and 9. We have seen one child in which the spinal cord had been completely severed by the ligaments at this point.

In the children with acute kinking, the cervical cord straightens itself spontaneously when the brain is removed from the cadaver, and thus the conventional method of removing the brain and cutting through the spinal cord from above means that the lesion could be completely missed. Some idea of the previous existence of such a kink can be obtained by handling the cervical spinal cord and medulla in the unfixed state after removal from the body, for if a normal cervical cord is pushed against the medulla, no acute kink or buckle will occur. On the other hand, when this is done with a child in which the kink has previously been produced and undone, the kink or buckle can be reproduced with relative ease. This does not apply after fixation in formalin and is no substitute for the direct inspection of the base of the skull during necropsy.

It is not the purpose of this paper to discuss the causes of acute swelling of the brain in children, but it must be stressed that it is a condition that is much more frequent than is usually recognized when a necropsy is carried out using conventional techniques.

There is little doubt that the compression of the upper cervical cord in these children does produce clinical symptoms and these may well be a major factor in producing death. The cases presented here were three children in whom the lesion was obviously of very recent origin. In the children with meningo-myelocele and hydrocephalus the kinking is obviously one of long standing and part of a very complex system of deformities, but the situation in cases 4 and 5, in which the kinking of the brain-stem appeared to be of long standing and occurring as a relatively isolated deformity, seem to form an intermediate group and can be considered a forme fruste of the Chiari type II deformity. In these two cases it is not possible to be sure of the cause of the lesion but both had symptoms and histories of phases of probable increase in intracranial pressure that could well have produced a situation as was found in cases 1, 2, and 3. More than that is pure speculation.

SUMMARY

Three cases of severe backward kinking of the brain-stem in the upper cervical region are recorded in children dying with an acute swelling of the brain. A similar but more chronic lesion was found in two children whose history suggested that they had had a previous phase of acute swelling of the brain.

The kinking of the brain-stem is similar but of less degree than that seen in the spur deformity of the Arnold-Chiari deformity.

This lesion appears to be due to the caudal displacement of the brain-stem beyond the limits permitted by the dentate ligaments.

The photographs are the work of Mr. A. Tunstall.

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