PLATYBASIA : WITH CASE REPORT

BY

CLARA MAYERSZKY

From the Neuropsychiatric University Clinic,* Debrecen, Hungary

From both pathological-anatomical and clinical aspects the condition called *impressio basilaris cranii*, "basilar impression," or "basilar invagination," was well known in the last century. In the last fifteen years it is referred to more often as "platybasia," in which the "basal angle" (the angle between the lines through the sphenoidal planum and clivus) is increased beyond the normal 135–140°.

Rokitansky (1846) is usually recorded as having first described it without any special name, but Berg and Retzius (1855) described it under the name *impressio baseos crani*. The first clinical observation dates from Boogaard (1865). Later it was described by Virchow (1857), Grawitz (1880), Schiffner (1878), Homén (1901), Krause (1911), and others. From the radiological point of view, Schüller (1911) had called attention to the condition. As Chamberlain (1939) had described four cases of platybasia, two of which had been successfully operated on by Temple Fay, it attracted attention in America. The term "platybasia," originally Virchow's, was introduced into the English-speaking literature by Chamberlain. Five cases were reported by Gustafson and Oldberg (1940), with a review of the whole subject. They found a connexion between the malformations around the foramen magnum and platybasia, and postulated that craniovertebral malformations have some relation to the hydrodilatation of the central nervous system (hydrocephalus, hydromyelia). Seven neurological cases with several malformations of the occipital bone, atlas, and axis were described by List (1941). His report discusses thoroughly the pathogenesis with regard to the ontogenetic factors, the mechanism producing the clinical picture, the diagnosis, and the treatment. Blomquist (1944) gives a detailed review of the whole question of the "basilar impressio," and data from the literature with reference to a case of his own, diagnosed post mortem. The only shortcoming in his paper is that he does not even mention the possibility of surgical treatment, although it had

come rather to the fore in the literature of the past fifteen years.

Primary Basilar Impression.—According to present-day opinion, basilar impression may be primary or secondary. Primary basilar impression is the result of a defect in craniovertebral development arising, according to Virchow's and Grawitz's suggestions, from the early ossification of the spheno-occipital suture, a defect in chondrogenesis and ossification. The posterior fossa fails to keep up in developmental pace with the vertebrae; the atlas overgrows the foramen magnum and does not join the condyles but fuses with the undermost part of the clivus and with the posterior rim of the foramen magnum. Schiffner suggested that the skull weighs heavily upon the atlas during intra-uterine life, so that the atlas fuses with the occipital bone and penetrates into the cranium. Becker pointed out in a thorough work that the basilar impression and the fusion of the atlas were to be considered as malformations and neither as variants of development nor as a caudal-progressive transformation leading to a future form of mankind, as suggested by others to account for the fusion of the atlas. He emphasizes that the fusion of the atlas and the basilar impression are in co-ordination and not in subordination, as they do not depend on each other in any genetic sense; both are determined in the third or fourth embryonal week. The primary idiopathic basilar impression is to be considered as a disorder of development, a view well supported by cases of basilar impression with other associated defects of development, such as, for example, spina bifida (List, 1941). Peyton and Peterson (1942) emphasize that the cases of basilar impression considered as defects of development are always accompanied by fusion to a varying degree of the atlas and occipital bone.

Secondary Basilar Impression.—This may occur in any diseases causing softening of the bones (Paget's disease, rickets, osteomalacia, hyperparathyroidism, lipoidosis, caries, senile osteoporosis, osteitis deformans, etc.). Owing to the plasticity of the basis, the uppermost cervical vertebrae—

*Chief, Prof. Dr. K. Sántha.

heavily pressed upon by the skull—are forced into the foramen magnum and the posterior fossa (Brailsford, 1944). Such cases of secondary platybasia are recorded by Schüller (1911), von Recklinghausen (1897), Homén (1901), Stilling (1890), Catola (quoted by Tscheslog), Kecht (1932), Tscheslog (1940), Antoni (quoted by Tscheslog), and Wycis (1944). The atlanto-occipital fusion is not a necessary concomitant of the secondary basilar impression.

Clinical and other Features

Skeletal Signs.—In the symptomatology there are some skeletal signs which provoke no particular complaints, such as the striking short neck, a certain restriction in movement of the head, the low hair-line at the back of the neck. These signs may increase in time, but not necessarily. The head may sink between the shoulders and become almost fixed. The basilar impression may assume clinical importance by causing neurological signs of direct compression on the pons and medulla oblongata, as in Bézi's case (1931) where the direct compression by the odontoid process caused softening in the medulla oblongata; or by the herniation of the cerebellar tonsils; or by hydrocephalus and hydro-myelia due to disturbances of the intracranial hydrodynamics, and occasionally by simultaneous defect of development of the central nervous system (for example, syringomyelia). From the point of view of symptomatology the cases of marked basilar impression of Stenvers (1916) and of Merio and Risak (1934), which were dominated by endocrine disturbances, are of some interest. In Stenvers' case of adiposogenital dystrophy there was found at post-mortem examination a macroscopically and microscopically normal pituitary body, but the third ventricle and the infundibulum had been dilated so enormously by the hydrocephalus that there was almost no basal wall of the ventricle, so that the author considered the clinical picture to be the result of the destruction of the autonomic centres. In one of their cases Merio and Risak considered eunuchoidism to be the effect of a lesion of the pituitary body due indirectly to basilar impression or to the internal hydrocephalus resulting from the basilar impression. In two other cases of theirs myxoedema was ascribed to disturbances of the circulation in the thyroid gland due to the very markedly lordotic cervical spine.

Neurological Signs.—The neurological signs (which may never be evident in cases of primary basilar impression during the whole life) develop gradually as a rule (Baruch, 1932; Tscheslog, 1940; Gustafson and Oldberg, 1940; Walsh and others,

1941; Wycis, 1944; etc.), although there are cases recorded with signs of rapid onset (Becker, 1940; Bodechtel and Guizetti, 1933). The symptoms may vary; the most striking of them are paraparesis, eventually quadriplegia, ataxia, nystagmus, disturbances of swallowing, difficulties of articulation, dyspnoea. In cases of secondary basilar impression the primary disease (for example, Paget's disease) may present many additional symptoms.

Radiology.—The x-ray investigation of the skull establishes the diagnosis. On the x-ray films in the lateral view the basal angle is increased to more than the normal 140° (though some authors allow 150° as the normal), the clivus approaches the horizontal line in different degrees; the shadows of the atlas and the odontoid process are above the so-called Chamberlain's line (the line between the posterior rim of the foramen magnum and the posterior edge of the hard palate), though normally they are below it. The foramen magnum is usually pressed upwards into the posterior fossa, and is narrowed, eccentric, and abnormal in shape. In cases of congenital origin the posterior arch of the atlas is usually fused with the lower edge of the occipital bone, and the odontoid process may reach over the foramen magnum. The spinous processes of the upper cervical vertebrae may be fused with each other. Encephalography usually shows a varying degree of dilatation of the ventricles.

Differential Diagnosis.—In the differential diagnosis cervical syringomyelia (Gustafson and Oldberg), chronic hydrocephalus (*ibid*), cerebellar tumour (Bodechtel and Guizetti), bulbar encephalitis (*ibid*), angle tumour (Adam-Falkiewiczowa and Nowicki, 1931), etc., must be considered. Gustafson and Oldberg suggest that in each case of cervical syringomyelia and in some cases of chronic hydrocephalus careful x-ray examination of the upper cervical spine, the foramen magnum, and the base of the skull should be undertaken.

Treatment

Treatment may be exclusively surgical: this, however, has been practised only in the last fifteen years. The patient whose case is recorded by Juhlin-Dannfelt (1933) died some weeks after operation. One of the four cases reported by Ebenius (1934) died while being operated on; three of them had slightly improved after surgical treatment. Two of the four cases recorded by Chamberlain (1939) had been operated on successfully by Temple Fay. In the same year at the International Congress of Neurologists in Copenhagen surgical treatment was suggested by Oljenick

he advised freeing the cerebellar tonsils which had been forced into the cervical canal from adhesions, and by this means bringing them back into the skull. In 1940 Gustafson and Oldberg described five patients who were operated on; two of them died. Dereymaeker (1941) described one patient who was slightly improved after surgical treatment. Of the six surgically-treated patients recorded by List (1941), three died of respiratory paralysis. The first of List's patients had been operated on in 1935 by Kahn. The cases recorded by Walsh, Camp, and Craig had been successfully operated on by the latter in 1941. Recently (1944) Wycis operated with excellent results on a case of basilar impression due to Paget's disease.

The surgical treatment consists essentially of upper cervical laminectomy with suboccipital decompression. List and Wycis suggested that the dura be left open, but Craig also had a good result with closure of the dura.

The following case is considered to be worth recording because the patient, who had severe symptoms, became practically free from complaints after operation and there are only a few instances reported of cases which have been operated on successfully.

Report of a Case

A farmer, aged 17, was admitted to hospital on Sept. 5, 1946, having been referred by his parents.

History.—Increasingly for one and a half years he had had headaches of the vertex, dull in character and most marked in the morning. He had also experienced occasional dizziness, staggering when working, and several attacks of nausea. For over a year swallowing had become difficult, and at the time of his admission he could not swallow fluid because it regurgitated through the nose. In the eight months before admission his speech had become gradually nasal and mumbling. Over the same period his left upper limb, then the left lower, later the right upper, then the right lower limbs had become clumsy and weak. He had been unable to walk for seven months and had been lying on his back or on his abdomen; he could not sit up by himself, and if he had been lifted up he had dizziness and dyspnoea. One month before admission the left side of his face became congested, but this settled down again. Since that time the sight of his left eye had been poor. He suffered from constipation and incomplete emptying of the bladder. His condition had not changed in the last four months. His parents had not taken him to the clinic because their doctor told them it was not worth while, that he would die anyway.

Family and past histories were irrelevant.

Examination.—He was an asthenic short boy rather undeveloped for his age. The skull was normal in size and form, and was painful to percussion. The neck

was short and markedly limited in all movements. There was on the ribs the so-called "rosary" of rachitic origin. There was marked lumbar lordosis, and marked dyspnoea on sitting up. The pulse rate was 76 per minute, the blood pressure 150/75 mm. Hg. The senses of smell and sight were good, and visual fields and fundi were normal. The pupils were medium-sized and circular; they reacted well to light and accommodation. The left pupil and palpebral fissure were smaller than the right. External ocular movements were full. There was a coarse, irregular, horizontal nystagmus when the patient looked to the right, and a finer one when he looked to the left. The trigeminal points were painful to pressure. There was bilateral constant wrinkling of the forehead which could be voluntarily abolished. The soft palate showed definite paresis on the left side with absence of the palatal reflexes on both sides. When he swallowed, the fluid came back through his nose. Speech was nasal. The tongue deviated to the left. His movements were slow; there was a spastic quadripareisis;

FIG. 1.—Diagram showing the increased basal angle.

he could not sit up, stand up, or walk, but there remained some motor powers in the extremities. The upper limbs were in the position of flexion, both lower limbs in extension. The fingers were in slight flexion, especially on the left hand; their movements were clumsy, weak, and limited. There was very marked atrophy in the muscles of both upper limbs, especially in the shoulder, thenar, hypothenar, and interossei muscles. Fasciculations sometimes occurred, particularly on the left side of the body. There was static and kinetic ataxia in the upper and lower extremities. The deep reflexes were markedly exaggerated, with knee and ankle clonus on both sides. On both sides plantar reflexes were extensor and abdominal reflexes and Meyer's and Léri's signs very sluggish. There was no sensory disturbance, but partial bladder and total bowel retention.

We could not obtain any cerebrospinal fluid by lumbar puncture. By cisternal puncture only one or two drops were obtained, even with suction.

A provisional diagnosis was made of syringomyelia in the upper cervical region.

X-ray examination clarified the diagnosis. The basal angle was increased to 160° (Fig 1). The arch of the

atlas was well delineated; it was not fused with the occipital bone, but they were nearer than usual to each other. The foramen magnum was asymmetric, flattened in the antero-posterior dimension, and narrowed. The upper cervical spine was markedly lordotic. Ventriculography was performed to complete the examination. There was moderate asymmetric hydrocephalus.

Operation.—On Oct. 17, 1946, the patient was operated on by Prof. Dr. K. Sántha. The patient having been placed in the face-down "cerebellar" position, under local anaesthesia, the surroundings of the foramen magnum and the posterior arches of the atlas and axis were exposed through a mid-line incision extending from the external occipital protuberance to the seventh

FIG. 2.—Diagram showing the situation at operation

cervical vertebra. The spinous processes of the third and fourth vertebrae were exposed only to get better access to the arch of the axis. The lower part of the occiput showed a flat indentation in the mid-line. The posterior arch of the atlas was large but smooth; there was no spinous process on it. The spinous process of the axis was large and bifid on the end; the arch was wide. The spinous processes of the third and fourth vertebrae were normal. The distance between the atlas and axis was normal but the atlas and occipital bones were nearer to each other than usual, although they were not fused.

The posterior arches of the axis and atlas were removed and then from a burr hole on the right side of the occipital bone the rim of the foramen magnum was

removed as well. The posterior rim of the foramen magnum was seen to extend far forwards below the cerebellar hemispheres so as to compress the dura with the effect of nipping the underlying tonsils. In the mid-line a huge bony comb was seen between the two cerebellar hemispheres, so that in order to progress we had to make a burr hole on the other side also and from there remove the posterior rim of the foramen magnum on the left side. At the end the thick part in the mid-line was pinched off with rongeur. The median bony comb was seen to extend far upwards and to rise inwards for over a half inch. The bone surrounding the foramen magnum was removed in a semicircle about one and a half inches high, and the comb rather more than this inwards. After opening the dura the arachnoid was seen to be normal, and a large amount of cerebrospinal fluid was obtained by opening it. The two cerebellar tonsils surrounded the bulbospinal border as flat structures and extended into the spinal canal to the level of the axis. The lower pole of the inferior vermis reached to the rim of the foramen magnum (Fig. 2). The tonsils, which were fixed only by some arachnoidal filaments, were freed and their poles lifted; only then was it possible to see the inferior cerebellar arteries, which had slipped down. We easily separated the tonsils from each other. The dura was left open as a decompression; the muscles and ligaments were closed with interrupted catgut sutures and the skin with interrupted black silk sutures. At the very beginning of the operation the patient had suffered respiratory paralysis, but this recovered and during the operation his condition was good.

Postoperative Period.—The neurological signs improved gradually, so that at his discharge five weeks after operation he was walking without any help, though the gait was still unsteady. There was slight ataxia in the finger-to-nose and heel-to-knee tests, and coarse nystagmus on looking to either side. No other signs, not even the marked lumbar lordosis, were seen.

Five months after operation he came alone by train for a follow-up examination. He stated that he had been working at home as a farmer. His one complaint was that he was not quite steady, particularly when walking in the dark. There was slow, coarse nystagmus on both sides. Gait was slightly ataxic but he was able to walk even with closed eyes. There were no other pathological signs. Of the former marked atrophy in the hand muscles there was nothing to be seen, and the muscles were well developed. He was able to turn his head to either side quite quickly and well, to bend forward on his chest; bending backwards was rather painful.

Comment

This case is considered to be a secondary platybasia of rachitic origin. At operation there were no signs of any developmental malformations of the atlas or of the cervical vertebrae; this supports the diagnosis. The rapid improvement of the neurological signs after operation confirms that there may be no malformations of the central nervous system such as syringomyelia. The (bulbospinal) neuro-

logical signs are perfectly explained by the findings at operation.

We should like to call attention to the fact that in some of the cases of primary platybasia there is only a limited degree of recovery achieved by decompression—a fact which is supported by some reports in the literature. Presumably in these cases we are faced with developmental malformations not only of the skeleton but of the central nervous system as well, and in these cases the simple decompression does not solve the problem. In the cases of obviously primary platybasia it is to be recommended that the exposed part of the spinal cord be examined very carefully at operation, and if there is a syringomyelia probably Puusepp's operation may be carried out at the same time as the decompression. Most of the cases in the literature with no satisfactory improvement after surgical treatment appear from the descriptions to be primary, and indeed in none of these cases is there record of any skeletal diseases as an aetiological factor.

Our similar case is a woman of 24 who, some months before the former case, was admitted to hospital with severe ataxia, four years' progressive quadriparesis, astereognosis in both hands, and hypoesthesia below the third cervical dermatome. High cervical laminectomy and suboccipital decompression (Prof. Sántha) produced only slight improvement in the neurological signs. She had complete occipitalization (fusion) of the atlas. We could not find any signs indicating a secondary origin of the platybasia, so it was considered to be a developmental malformation. At operation the cervical spine did not show any signs of syringomyelia; it is possible, however, that this patient did not recover sufficiently because she had some additional developmental malformation of the central nervous system.

As surgical treatment is the only possible therapy, it should be advised in every case, even in the cases of undoubtedly primary origin. We may hope for complete recovery of the neurological signs, not only in secondary but in primary cases (Walsh and others, 1941).

Summary

1. The literature regarding platybasia is reviewed.

2. A case of secondary platybasia of rachitic origin is described which was relieved by upper cervical laminectomy and suboccipital decompression.

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