PLATYBASIA

REPORT OF TEN CASES WITH COMMENTS ON FAMILIAL TENDENCY, A SPECIAL DIAGNOSTIC SIGN, AND THE END RESULTS OF OPERATION*

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PLATYBASIA, also referred to as basilar impression or invagination, has been well described by many authorities.[1-10] It is easily diagnosed by roentgen ray yet often missed simply because it is not looked for. It is frequently mistaken for multiple sclerosis and syringomyelia. It is the purpose of this paper to present ten cases of platybasia with and without concomitant Arnold-Chiari and syringomyelic defects and to comment on its prevalence and the surgical results. Proof of a familial tendency is offered and a special diagnostic sign is stressed.

Peyton and Peterson,[1] Gustafson and Oldberg[2] and Hadley[3] have presented excellent reviews of the literature. A fundamental feature of platybasia is the deformity of the foramen magnum and the invagination of the cervical spine into the cranial cavity, so much so that occasionally the first cervical vertebra is hidden within the foramen magnum (see Case 4). In the majority of cases this is associated with other congenital anomalies of the skeleton and brain stem, especially an Arnold-Chiari deformity and syringomyelia. Occasional cases of platybasia have followed Paget's disease or osteogenesis imperfecta.[4, 6] Although congenital in origin, symptoms generally begin in the third to fifth decade of life, with the two extremes of four years and 79 years of age having been reported.

Clinically, platybasia may simulate multiple sclerosis, syringomyelia, other degenerative cord diseases, and hydrocephalus. Compression of the contents of the posterior fossa with angulation of the medulla over the intruding odontoid process produces nystagmus, staggering gait and sensory loss, which is often associated with pyramidal tract signs. There may be a slow progression to complete spastic quadriplegia, various cranial nerve palsies, bizarre respiratory patterns and a dry cough. The adhesions forming about the herniated cerebellar tonsils may cause hydrocephalus. Patients usually exhibit a short, squat neck with backward tilting of the head. Diagnosis is confirmed roentgenographically both by a distorted, narrow foramen magnum in the basal view and by the elevation of the odontoid process above Chamberlain's line[7] in the lateral view, although this may occur to a slight degree in 5 per cent of normal patients.[8] The atlas is generally fused to the occiput, as shown by roentgenogram in the flexed position,[3] and may even be hidden within the foramen magnum. An accompanying Arnold-Chiari deformity may be diagnosed by myelography.[11]

The only known effective treatment is surgical decompression of the foramen magnum and the upper cervical spine. The dura should be opened in order to free up the adhesions and decompress the Arnold Chiari malformation. Certain surgeons amputate the cerebellar elongated tonsils.

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CASE REPORTS

Case 1.—Coarse nystagmus with eyes in neutral position; accompanying Arnold-Chiari deformity and club feet. There was permanent arrest of progression without improvement.

A 32-year-old white male was admitted to the Hartford Hospital in June 1942 with congenital bilaterally clubbed feet. There had been onset 2 years previously of a severe, atypical, coarse nystagmus occurring vertically with the eyes in neutral position but horizontally upon either lateral position. There was progressive numbness and astereognosis of right hand with ataxia of right arm and both legs and hypesthesia to pain in the right hand. A squat neck was noted and there were normal spinal dynamics and protein. Roentgen ray confirmed the presence of a definite platybasia with incomplete development of the atlas. Suboccipital decompression was carried out with the removal of the first and second cervical laminae, exposing an Arnold-Chiari deformity with cerebellar tonsils extending to the upper border of C3 vertebra surrounded by marked adhesions. Convalescence was complicated by sterile meningitis postoperatively. The patient has shown no change in his neurologic status over the ensuing eight years, in contradistinction to a rapid progression preoperatively.

Case 2.—Mistaken diagnosis of multiple sclerosis in two medical centers; nystagmus; slowly progressive quadriplegia. There was xanthochromic fluid and accompanying Arnold-Chiari malformation. Mild improvement following operation.

A 44-year-old white female was admitted to the Hartford Hospital in June 1946 with a progressive spastic quadriparesis, legs worse than arms, over preceding 5 years. Chair-ridden for 2 years. Horizontal nystagmus to the left with bilateral pyramidal tract signs. Xanthochromic fluid with a high protein of 60 mg. per 100 cc., although 6 months previously pantopaque had flowed freely into the pontine cistern. Operative decompression in June, 1946, revealed a herniation of the cerebellar tonsils to mid-C2 vertebra, with dense adhesions at the level of the foramen magnum. Adhesions were lysed, necessitating resection of a tortuous redundant posterior-inferior cerebellar artery. There was postoperative decrease in spasticity and increase in motor function of the upper extremities. Spinal fluid dynamics and protein returned to normal in 3 months' time.

Case 3.—Hydrocephalus with papilledema; six months' cure; recurrence with mental deterioration and death one year later.

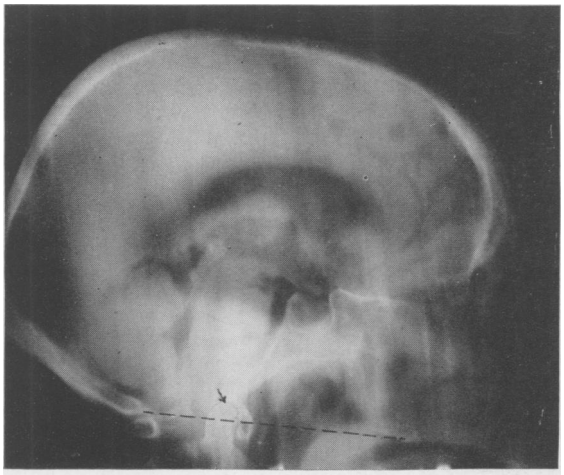

FIG. 1

FIG. 1.—(Case 10). Lateral laminograph to show elevation of odons (arrow above Chamberlain's line (dotted).

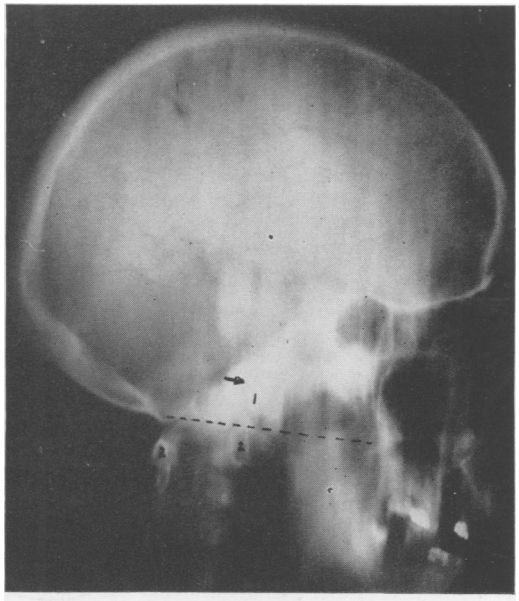

FIG. 2

FIG. 2.—(Case 4). Lateral laminograph to show atlas (1) hidden within the foramen magnum. Axis (2) is easily mistaken for the atlas. Arrow marks extreme elevation of odons above Chamberlain's line (dotted).

A 64-year-old woman was admitted to Hartford Hospital in June, 1946, with a very short neck, initially disabled 2 years previously by weakness of left shoulder and biceps muscles, right-sided hypesthesia below clavicle, progressive headaches, visual failure and finally choked disks with hemorrhages. A ventriculogram showed a symmetrical hydrocephalus with mild herniation of cerebellar tonsils and a spinal fluid pressure of 230 and protein of 61 mg. per 100 cc. Roentgen rays revealed a platybasia. Operative decompression was done in June, 1946. The atlas was invaginated into the foramen magnum; no adhesions. Postoperative pneumoencephalogram revealed recession of the hydrocephalus. Six months later there was recurrence of papilledema, increased intracranial pressure to 490 mm. of water and hydrocephalus. There was a free flow between ventricular and spinal subarachnoid systems. Over the ensuing months she underwent bilateral subtemporal decompressions and finally choroid plexectomies. This cured her increased pressure but not the gross mental deterioration. She died in a nursing home in May, 1947. Autopsy was refused .

Case 4.—(Sister of patient in Case 5). Headaches, dizziness, staggering gait, familial history accompanying Arnold-Chiari deformity, coarse nystagmus. Symptomatic cure was achieved but little change in neurologic status. There was no further progression.

A 33-year-old white female was admitted to the Hartford Hospital in April, 1947, with episodic headaches and dizziness for previous year, mildly progressive ataxic gait, short squat neck, coarse nystagmus on looking to the right, and absence of left abdominal reflexes. Roentgen rays revealed platybasia with fusion of the atlas to the occiput. Spinal puncture was normal. Pneumoencephalogram failed to fill the ventricular system. In April, 1947, a suboccipital decompression was performed, with the findings that the atlas was invaginated into the foramen magnum, with elongation of the cerebellar tonsils down to the top of the second cervical vertebra, and fine arachnoidal adhesions surrounding the tips of the tonsils. The adhesions were freed. Postoperative severe headaches subsiding in one month. Neurologic conditions were unchanged by the operation, except for complete

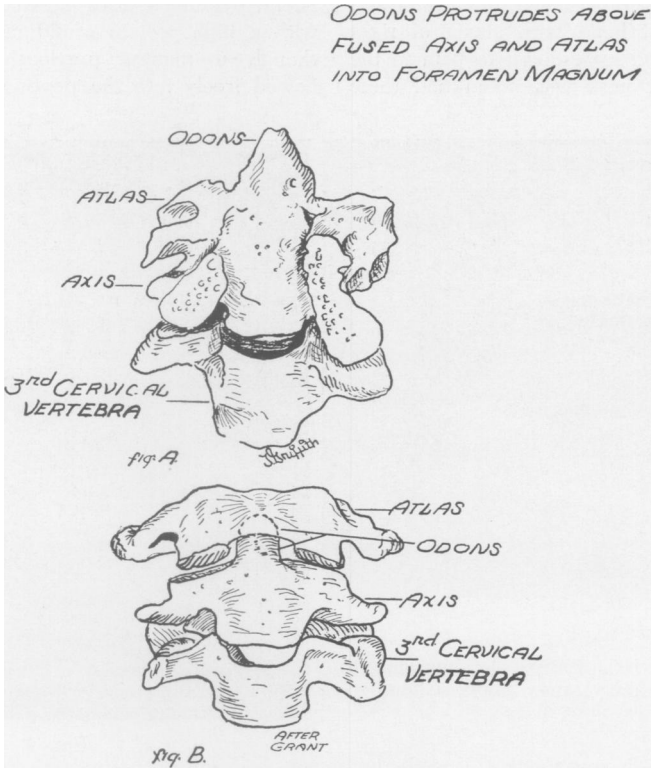

FIG. 3.—(Case 8). Autopsy specimen of upper cervical spine to show gross anomalies. Sketch illustrates comparison with the normal cervical spine (ventral view).

subsidence of headaches, dizziness and questionable improvement in her staggering.

Case 5.—(Sister of patient in Case 4, with identical clinical picture.) There was no improvement following operation, and no further progression.

Case 6.—Ataxia, mild paraparesis, and coarse horizontal nystagmus were present. There was possible familial history. Accompanying was an Arnold-Chiari deformity. No improvement following operation, and there was no further progression.

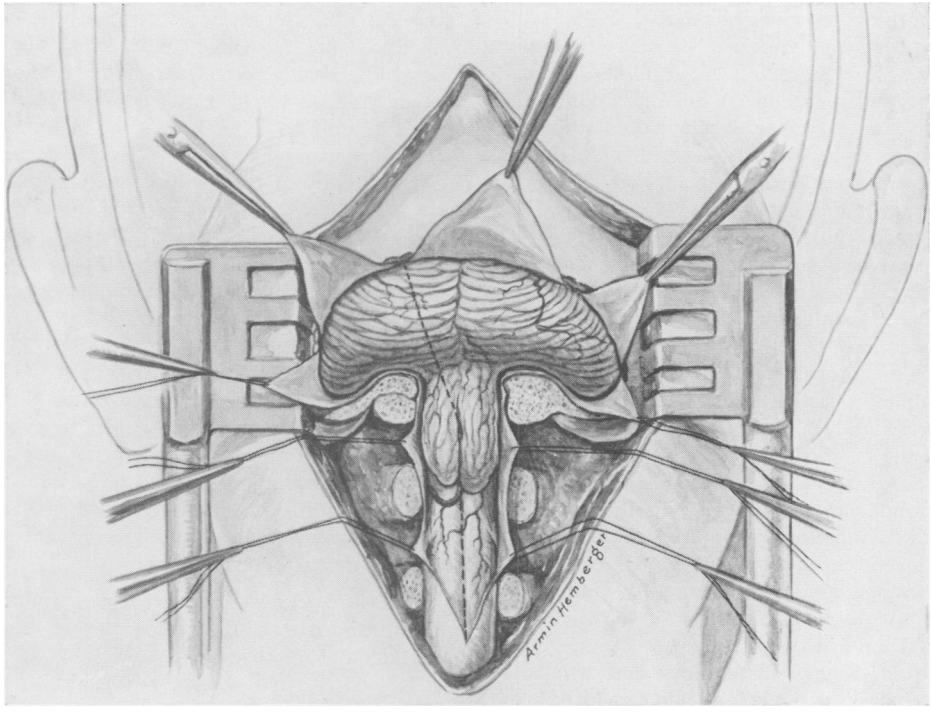

FIG. 4.—Composite sketch to illustrate operative exposure with partially fused occipito-atlas bones and accompanying Arnold-Chiari defect. Dotted line indicates line of dural incision.

A 48-year-old white female was admitted to the Hartford Hospital in November, 1947, with identical but more severe signs and symptoms than those found in the patient of Case 4. She had an ataxic gait on a broad base for the previous 14 years, with no progression before or after operation. Pernicious anemia tests were negative at another hospital. Mild hypertension of 180/96. There was a short squat neck, with increased forward angulation at cervico-dorsal junction, and lateral nystagmus on bilateral gaze. There were increased deep reflexes in the legs without Babinski, and fizrillations of the tongue and bilaterally depressed corneal reflexes. Roentgen rays revealed platybasia, with bony fusion between the axia, atlas and occiput. Decompression was carried out in November, 1947, with finding of the atlas invaginated well into the foramen magnum and elongation of the cerebellar tonsils to the level of the second cervical vertebra. There were no adhesions. Convalescence was uneventful. Neurologic status was unchanged following the operation over a two and three-quarter year follow-up period.

This was a 31-year-old single, white female with a 10-year history of slight ataxic gait and slight weakness of legs, very slowly progressive, with coarse horizontal nystagmus with eyes in neutral position and when looking upwards, worse on looking to the right. There was diminution of the right corneal reflex, deviation to the right in gait. Roentgen rays revealed platybasia with fusion of the atlas to the foramen magnum. Her mother has a short neck and unsteadiness of gait although roentgen rays show no gross evidence of platybasia. Decompression done in June, 1947; elongation of cerebellar tonsils down to the inferior edge of C2 vertebra. There were no adhesions. Normal convalescence followed and there was no further progression or improvement postoperatively.

Case 7.—There were atypical rotary nystagmus, unequal pupils, and bilateral pyramidal tract signs, with accompanying syringomyelia and Arnold-Chiari deformity. A nephew has high cervical syringomyelia and possible Arnold-Chiari syndrome (Supplementary Case 7).

A 30-year-old elevator man was admitted to the Hartford Hospital in March, 1948, with a slowly progressive weakness of the right arm and leg over the preceding 6 years, with loss of sweating and sensation on the right side of body over 2 years. Neurologic examination revealed an atypical rotary nystagmus, especially on right lateral gaze; hypesthesia for pain, loss of sweating and vasodilatation of arm; and asymmetrical pyramidal tract signs. Pneumo-encephalogram failed to fill the ventricles with normal spinal dynamics and protein. Roentgenograms showed borderline platybasia. Pantopaque myelogram revealed a scalloped defect[10] at mid-C2 vertebra, indicating an Arnold-Chiari anomaly as well as a platybasia. Suboccipital decompression was performed. The dura was tightly filled by an Arnold-Chiari extension of the cerebeller tonsils down to mid-C2 vertebral level, with dense adhesions surrounding the tonsils and completely occluding the foramen of Magendi. Adhesions were freed. Convalescence was uneventful, and there was no gross improvement postoperatively, but also no further progression of his disability. Because of the slight fibrillations at the first dorsal interosseous muscle of the right hand, segmental and dissociated pain and temperature loss, diminished reflexes in right arm and autonomic changes, this man probably has a syringomyelia as well as his other anomalies.

Supplementary Case 7.—(Nephew of patient in Case 7.) Scoliosis, bilateral pyramidal tract signs, absent arm reflexes, segmental loss of pain and temperature. No operation was performed.

A 25-year-old nephew of the patient in Case 7 reported because of familial tendency, with a clinical picture of syringobulbia manifested by an old scoliosis, weakness in one leg, bilateral Babinski, increased leg reflexes, absent arm reflexes, segmental pain and temperature loss, D1-D5 dermatomes on the left side and below the nipple line on the right side and weakness of the left hand. Roentgen rays revealed odons extending up to but not over Chamberlain's line. No operation was performed.

Case 8.—Patient showed spastic quadriparesis, nystagmus, extreme platybasia, Klippel-Feil and cervical rib anomalies. Operation was followed by death from old and recent intramedullary cord hemorrhage, probably secondary to pressure from odons.

A 49-year-old man was admitted to Hartford Hospital July, 1949, with neck pains and numbness of fingers for years, progressive spastic quadriplegia over the previous year, pyramidal tract signs, ataxia, vibratory loss, fine nystagmus, short squat tilted neck, extreme platybasia, Klippel-Feil fusion of first and second cervical vertebrae and bilateral cervical ribs. A pneumo-encephalogram failed to fill the ventricles; spinal fluid protein was 41 mg. per 100 cc. with normal dynamics. A suboccipital decompression was performed under intratracheal ether anesthesia, with the patient in upright position with head only slightly flexed. Cisterna magna obliterated by cerebellar tonsils without herniation. No adhesions were present. The dura was left open, with arachnoid intact. The operation went smoothly, but immediately postoperatively the patient developed gasping respirations, with pulmonary edema and respiratory failure, and death followed. Autopsy revealed extreme platybasia (Fig. 3), with fusion of atlas and axis and defective odons protruding 1.5 cm. above the top of atlas and well into the foramen magnum, with direct pressure against and angulation of the ventral spinal cord at the junction of the medulla and cervical cord. At this level the cord contained an extensive old intramedullary hemorrhage and was thinned out to a mere ribbon.

Case 9.—There was progressive spastic paraparesis and nystagmus. Decompression resulted in considerable improvement.

A 43-year-old woman admitted to the Hartford Hospital in October, 1948, with progressive weakness and stiffness of legs and subjective weakness of the right arm over one and one-half years, progressing to an inability to walk more than a few steps. There was nystagmus on right lateral gaze. Roentgen ray showed the odons 5 mm. above Chamberlain's line. A suboccipital decompression revealed upward invagination of the foramen magnum with the cerebellar tonsils extending 1.5 cm. below the foramen and exhibiting a marked dorsal indentation by the posterior rim of the foramen. Rapid improvement occurred postoperatively, with ability to walk several blocks and perform housework, continuing to date.

Case 10.—Burning pain in neck and arms since episode of coughing 7 years previously; right-sided weakness and numbness. Concomitant syringomyelia. The patient improved with operation.

A 42-year-old woman was admitted to Hartford Hospital January, 1950, because of burning pain in neck and arms progressive since a coughing episode 7 years previouly. There was progressive mild weakness of right arm and leg, with numbness of the arm and face; muscular twitchings of

right arm; loss of temperature sense on right with segmental right-sided sensory loss of cheek and from the second cervical to the third dorsal dermatome; and weakness of the right trapezius muscle. There were normal spinal fluid dynamics and protein of 31 mg. per 100 cc. There was a negative myelogram. Roentgenograms revealed elevation of odons above Chamberlain's line (Fig. 1). Pneumo-encephalogram resulted in an absence of ventricular filling. Suboccipital decompression confirmed the presence of a platybasia and Arnold-Chiari syndrome and resulted in temporary relief from pain, with subsequent return to a lesser degree 4 months' postoperatively and a slow, slight improvement in the motor weakness over ensuing 6 months.

DISCUSSION AND CONCLUSIONS

These case presentations have been made in order to stress certain aspects of platybasia which have not heretofore been given sufficient prominence. Platybasia is more prevalent than is commonly known. All but three of these cases have occurred over a two-year period in the neurosurgical practice of one man. The diagnosis can readily be missed because the bony deformity is not looked for in routine films. Even when searched for, the atlas may be hidden within the foramen magnum, giving a false appearance of normality (Fig. 2). Eight of the ten cases were diagnosed multiple sclerosis or syringomyelia over long periods of time by competent neurologists. As far as these writers can ascertain, no previous mention has been made of a familial tendency. Two of the above cases occurred in sisters (Cases 4 and 5). One case with platybasia, Arnold-Chiari and syringomyelia, has a nephew with clinical syringobulbia and a possible platybasia and Arnold-Chiari syndrome (Cases 7 and supplementary 7). The mother of another patient (Case 6) has always had a short neck and mildly unsteady gait. All but one case had other congenital defects: an Arnold-Chiari (more accurately an Arnold) deformity in eight cases; syringomyelia in three; club feet in one and cervical ribs and Klippel Feil anomaly, each in one case. The age of onset of symptoms averaged 37 years and in all but two patients began after the age of 30 years. In addition to the neurologic signs of an ataxic gait, pyramidal tract signs, sensory changes and squat neck described in the literature, these authors wish to stress a special sign occurring in the majority of cases consisting of an atypical, coarse type of nystagmus frequently occurring with the eyes in neutral position and changing its character or direction depending on whether the eyes are turned laterally, upwards or at rest.

In diagnosis, myelography, as first described by one of the authors,[11] is of considerable aid in revealing the presence of constriction and an Arnold-Chiari deformity (Case 7). If the patient's head is kept hyperextended and under no circumstances rotated sideways nor the patient allowed to cough, 9 cc. of pantopaque may be run up the spine and through the foramen magnum onto the clivus for a single A-P spot film of the foramen magnum and then returned caudally without any residuum being left within the skull. Pneumo-encephalography is a dangerous but confirmatory diagnostic aid, exhibiting an absence of filling of both the ventricular and basal cisternal systems accompanied by a marked physical reaction (Cases 4, 7, 8 and 10).

Surgical decompression in this series gave relief from subjective pain and dizziness. It resulted in an arrest but only occasional improvement (three cases) in the neurologic disability. Therefore, operation should be performed early in cases showing progression, but is of doubtful value in stationary cases. The decompression should be carried out through a vertical incision with the patient upright or on his side. Beware of too much anterior flexion of the neck in an upright position because of compression of the medulla by the odons (Case 8). The dura is opened with two parallel vertical or V relaxation incisions in order that the dura may remain as a funnel-

shaped cradle for the elongated cerebellar tonsils. The tonsils are gently mobilized through an intact arachnoid in order to free them from adhesions. The authors have not amputated them in their desire to keep the arachnoid intact and because of the presence in all cases of an anomalous redundant posterior-inferior cerebellar artery which may completely traverse the dorsal surface of the tonsils. It is the authors' concept that the angulation of the medulla over the abnormally high odontoid process is the chief offender in causation of the neurologic signs and disability. Posterior decompression simply prevents adhesive arachnoiditis and hydrocephalus. Future surgical advance lies in the development of a successful removal of the odons itself, possibly from an anterior approach through the mouth,* thus preventing its impingement on the ventral surface of the cord and medulla.

SUMMARY

1. Case presentation is made of ten patients having platybasia. Nine had concomittant congenital anomalies of the cord, brain or skeleton.

2. Platybasia is more prevalent than heretofore known.

3. A familial tendency is reported.

4. The special diagnostic sign most frequently encountered is an atypical, coarse nystagmus, varying with the position of the eyes.

5. Surgical decompression will arrest progression but only occasionally improves the neurologic disability.

6. Details of diagnostic and surgical technics are described.

* This has been found feasible on a cadaver.

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