Basilar impression and Arnold-Chiari malformation

A study of 66 cases¹

M. CAETANO DE BARROS, W. FARIAS, L. ATAÍDE, AND S. LINS

From the Institute of Neurology and Neurosurgery, Medical School, Federal University of Pernambuco, Brazil

We have been interested for a long time in the problem of the deformation of the base of the skull known as basilar impression and described by the anatomists since 1790 (Ackermann), although only recently recognized from the clinical point of view. This condition has attracted our attention particularly because of its high incidence in the North-east of Brazil and has given rise to other papers written by one of us (de Barros, 1957, 1959).

The possibility of precise pre-operative diagnosis has been established by Chamberlain's paper (1939). This author described a line in plain radiograph of the skull which permits recognition of the malformation under discussion. Chamberlain's line is that which in a lateral view of the skull goes from the dorsal lip of the foramen magnum to the dorsal margin of the hard palate and in normal individuals passes above the atlas and the tip of the odontoid process. When a basilar impression is present, these structures are above this line.

Phillips (1955) considers as arbitrary any definition of basilar impression based on Chamberlain's line, although admitting that it exists when the tip of the odontoid process is found more than 5 mm above this line. To McRae (1953) the limit would be only 3 mm.

As it is not always easy to draw Chamberlain's line in a plain radiograph of the skull, one can resort to the tomograph or to other referential lines. McGregor's line (1948) connects the dorsal rim of the hard palate to the lowest point of the occipital bone. In normal individuals this line almost superimposes that of Chamberlain, whereas in people having basilar impression it tends to be separate from it. Bull (1946) and Bull, Nixon, and Pratt (1955) point out that the planes passing by the hard palate and by the atlas are parallel or they slightly intersect, this relation being altered when there is basilar impression (Fig. 1).

Fischgold, David, and Brégeat (1952) described two lines, both of them drawn in anteroposterior view of the skull. The first joining the tips of the mastoid processes usually passes at the level of the atlanto-occipital joints and to 3 mm above or below the tips of the odontoid process. In patients with basilar impression the atlanto-occipital joints and the odontoid process are clearly above this line. The second line drawn between the two digastric grooves passes well above the tip of the odontoid process in normal skulls (Fig. 2). As routine for the radiological diagnosis of basilar impression we take the Chamberlain's and Fischgold's lines taken in plain views or in tomographs of the skull (Fig. 3).

Basilar impression may be acquired, the deformation of the skull deriving from a systemic osseous disease like Paget's disease, osteoporosis, rickets, hyperparathyroidism, osteochondro-dystrophy, etc., or—as is more frequent—it may be primary, a congenital malformation.

We believe that in its primary form basilar impression derives from an embryonic disturbance. Such an assertion is supported by its frequent association with other anomalies of the skeleton such as morphological alteration of the foramen magnum (narrow, asymmetrical, shaped like hearts of playing cards), hypoplasia of the atlas, Klippel-Feil anomaly, spina bifida, etc.

A frequent operative finding is an association of basilar impression with a displacement of the odontoid process. Another frequent association is with the Arnold-Chiari malformation. This latter condition was classified by Chiari (1896) into three types:

I—Cerebellar tonsils and lower part of the medulla below the foramen magnum, without displacement of the 4th ventricle.

II—Caudal migration of the lower part of the cerebellum still more pronounced, associated with downward displacement of the 4th ventricle which appears lengthened and with its foramina opening into the spinal subarachnoid spaces.

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FIG. 1. Basal angle: AB—Chamberlain's line. CD—Bull's line.

FIG. 2. Basilar impression. Fischgold's lines: AB—Bi-digastric line. CD—Bi-mastoid line.

FIG. 3. Basilar impression. Lateral planigraphy: AB—Chamberlain's line.

III—Cerebellum and medulla displaced into the cervical spinal canal within a meningocele.

From these three types only those of type I and exceptionally some of type II reach adult age and are the subject of this paper.

Patients classified as belonging to type III and a great majority of those of type II do not survive, as has been well demonstrated by Russell (1949).

MATERIAL AND METHODS

Our material is represented by 66 patients examined in the Institute of Neurology and Neurosurgery, Medical School of the University of Pernambuco, Brazil, and in private practice until 1964. Since then we have operated upon 15 further cases, giving a total of 71 cases. Some of these have already been discussed in previous papers (de Barros, 1957; de Barros, Pernambucano, Hazin, Maia, and Ataide, 1957; de Barros, 1959).

In the present study we include only those patients whose clinical manifestations were caused by the malformations under discussion. We put to one side another group, perhaps larger, in which the diagnosis of basilar impression was suspected by the morphology of the head and confirmed by radiological studies but whose

very heterogeneous complaints had nothing to do with the osseous anomaly. All patients were born in the North-east of Brazil, 52 were male and 14 female (Table I). This marked preponderance of the male sex in our series could perhaps be partially explained by the fact that we dispose of a greater number of beds for men than for women in our wards.

TABLE I
SEX DISTRIBUTION

Sex	Group I	Group II
Male	25	27
Female	8	6

As to the racial type, 35 patients were white, 29 mulattoes and only two were negroes (Table II). The population of the North-east of Brazil is formed by a little more than 50% of mulattoes and about 2% of negroes, the rest being white. The lowest social and economic classes are predominantly represented by mulattoes and our Service admits only poor patients. Even so the majority of the cases in our series are white.

TABLE II
RACIAL TYPE

Racial Type	Group I	Group II
White	18	17
Mulatto	14	15
Negro	1	1

It should be noted that the North-east of Brazil had been occupied by the Dutch in the 17th century for 34 years and it has been said that basilar impression is a common condition in the Netherlands. It may be that there is some connexion between these facts.

The cases are separated into two equal groups of 33 patients. Group I consists of cases verified by surgery (32 cases) or by necropsy (one case). This group includes 22 cases of basilar impression associated with Arnold-Chiari malformation operated on, and one case verified by necropsy; seven cases of pure basilar impression operated on; three cases of pure Arnold-Chiari malformation also operated on.

Group II consists of patients with a diagnosis of basilar impression associated or not with Arnold-Chiari syndrome but for several reasons not operated on. Every patient, apart from the neurological examination, according to the routine of the Service, had a radiograph of the skull taken in four positions (P.A., lateral, Hirtz, and Towne). Some of them also had tomographs. The diagnosis was established through Fischgold's and Chamberlain's lines. For the latter we accepted 5 mm above this line as the maximum limit of normality for the tip of the odontoid.

Some patients had spinal fluid examinations carried out, always done by lumbar puncture. We never do suboccipital puncture in this sort of patient, since it may not succeed because of the blockade of the cisterna magna by the herniated cerebellar tonsils and also because of

the danger of bleeding from puncture of large veins that sometimes exist on the dorsal aspect of these tonsils, as we have quite often seen during operation. We recognize that the number of cases in which these anomalies were found separately is very small, but as already emphasized by one of us (de Barros, 1959) in a previous paper we have found it possible to make the pre-operative differential diagnosis on clinical grounds.

ANALYSIS OF THE MATERIAL AND COMMENTS

AGE We had no cases in the first decade, only two in the fifth, and four cases in the sixth decade. The greatest incidence was in the third decade (24 cases) followed by the second and fourth decades with 18 and 17 cases respectively (Table III).

TABLE III
AGE OF PATIENTS

Age (yr)	Group I	Group II
10-19	11	7
20-29	8	16
30-39	11	6
40-49	2	—
50-59	1	3

AGE OF ONSET OF SYMPTOMS The clinical history of these patients usually dates back several years, with insidious symptoms and an evolution needing a very careful inquiry to pinpoint the appearance of the same which, in the majority of cases, was already forgotten.

This makes it difficult to establish with certainty the age at which complaints begin. The analysis of Table IV permits us to place the appearance of the symptoms, in the great majority of cases, in the second and third decades (24 and 26 cases respectively). However, they may appear early in the first decade (five cases) or late (three cases in the sixth decade). We had seven patients whose complaints started in the fourth decade and in only one of our patients the complaints came from the fifth decade (Table IV).

TABLE IV
ONSET OF SYMPTOMS

Age (yr)	Group I	Group II
0-9	5	—
10-19	12	12
20-29	10	16
30-39	6	1
40-49	—	1
50-59	—	3

Attention is drawn to the relatively late onset, although, as it is a congenital anomaly, one would expect it to be revealed in early life. Explanation

could be found in a progressive ascension of a thinnish hypoplastic basisphenoid, as mentioned by List (1941, 1957) under the action of the counter pressure of the spine against the weight of the head associated with dislocation of the odontoid process and deformation of the foramen magnum.

Progressive summation of these factors would produce direct or indirect damage to the adjacent nervous structures. It seems obvious that these natural aggravating conditions may be further worsened by addition of other mechanical factors, for instance carrying weights on the head. This could perhaps be a partial explanation for the preponderance of the condition among males in our material.

We further mention the frequent finding of local reactive alterations of the meninges, represented by fibrous thickening and adhesive or cystic arachnoiditis, whose compressive effects add to those mentioned above. Furthermore, we have still to consider the possibility of aggravation being connected with disturbance of blood supply and disorders of the cerebrospinal fluid circulation.

The pre-existent nervous lesions (hypoplasias, degenerative alterations) may themselves become progressively worse, spontaneously or by the action of bone compression as List (1941, 1957) has already mentioned.

ANALYSIS OF SYMPTOMS

The analysis of the symptoms which caused the patients to seek medical examination discloses that the most common is weakness in the lower limbs (68%), followed by unsteadiness of gait (56%), and headache (53%). Paraesthesias in the form of formications, numbness, burning sensations, etc., usually localized in the upper limbs, appear in 43% of cases. Dizziness and dysphagia were mentioned by more than one-third (37%) of the patients.

Another very frequent complaint was pains in the nape of the neck (28%), of variable intensity in the majority of cases, precipitated by coughing, sneezing, straining, and even laughing. These pains are possibly connected with disturbance of the uppermost cervical roots (C2 and C3), compressed or stretched, especially when there is an Arnold-Chiari malformation associated with basilar impression.

Other symptoms less frequently mentioned were: sexual troubles, in the form of reduction of libido or even impotence (27%); nasal voice (27%); diplopia (22%); and dizziness (21%). Several other complaints are found less frequently (Table V).

A study of the symptoms in those patients in which there were pure forms of each anomaly seems to us of great interest. We have noticed that in the

TABLE V
GENERAL SYNOPTIC TABLE: SYMPTOMS
GROUP I-II

	Group I (no.)	(%)	Group II (no.)	(%)	Total (no.)	(%)
Pain in neck under strain	15	45	4	12	19	28
Unsteady gait	25	75	12	36	37	56
Diminished hearing	3	9	2	6	5	7
Dysphagia	14	42	11	33	25	37
Nasal reflux	6	18	1	3	7	10
Diplopia	9	27	6	18	15	22
Dysarthria	5	15	2	6	7	10
Dizziness	12	36	13	39	25	37
Weakness in limbs	20	60	25	75	45	68
Paraesthesia in face	6	18	1	3	7	10
" " limbs	19	57	10	30	29	43
Paracusia	5	15	5	15	10	15
Headache	19	57	16	48	35	53
Sphincter troubles	12	36	7	21	19	28
Sexual troubles	11	33	7	21	18	27
Diminished vision	4	12	1	3	5	7
Vertigo	10	30	4	12	14	21
Nuchal rigidity	3	9	—	—	3	4
Nasal voice	9	27	9	27	18	27
Vomiting	7	21	6	18	18	27
Blurring of vision	9	27	2	6	11	16
Irregular menses	2	6	—	—	2	3
Collapse of legs	1	3	1	3	2	3
Dyspnoea	2	6	—	—	2	3
Sleepiness	2	6	1	3	3	4
Seizures	2	6	—	—	2	3
Fainting	1	3	—	—	1	1

cases of basilar impression the dominant complaints were weakness and paraesthesias in the limbs (85%), whereas in the patients with the Arnold-Chiari syndrome the dominant symptom was unsteadiness of gait, which was present in every case. Furthermore, dizziness, pains in the nape of the neck, stiffening sensations and headaches, although often present in the cases of pure basilar impression, appear in a more significant manner in the cases of pure Arnold-Chiari syndrome.

In summary, a comparative study of symptoms of the patients with isolated forms of each anomaly has shown that, whereas in the basilar impression motor and sensory disturbances are more frequent, in the Arnold-Chiari syndrome the clinical picture is basically made of cerebellar and vestibular disturbances besides signs of involvement of the lower cranial nerves (Table VI).

As we have already said, the symptoms, in the great majority of cases, have a progressive course. Only two of our patients, both adolescents, had a sudden history beginning with headache, vomiting, and unsteadiness of gait. An association of basilar impression and Arnold-Chiari deformity was present in both. In these cases the syndrome of intracranial hypertension was due to cerebrospinal fluid block caused by herniated cerebellar tonsils.

ANALYSIS OF SIGNS AND SYMPTOMS

Mere inspection of the patients with basilar

TABLE VI

COMPARATIVE TABLE OF SYMPTOMS IN THE CASES
WITH B.I. OR A.C. ALONE AND B.I. + A.C.

Complaints	B.I. (7 cases)		A.C. (3 cases)		B.I. + A.C. (23 cases)	
	(no.)	(%)	(no.)	(%)	(no.)	(%)
Pain in neck under strain	3	42	2	66	10	43
Unsteady gait	3	42	3	100	19	82
Diminished hearing	1	14	—	—	2	8
Dysphagia	1	14	2	66	11	47
Nasal reflux	—	—	1	33	5	21
Diplopia	—	—	—	—	9	39
Dysarthria	—	—	—	—	5	21
Dizziness	3	42	2	66	7	30
Weakness in limbs	6	85	1	33	13	56
Paraesthesia in face	1	14	—	—	5	21
„ „ limbs	6	85	2	66	11	47
Paracusia	1	14	—	—	4	17
Headache	3	42	2	66	14	60
Sphincter troubles	4	57	—	—	8	34
Sexual troubles	2	28	—	—	9	39
Diminished vision	—	—	—	—	4	17
Vertigo	2	28	—	—	8	34
Nuchal rigidity	—	—	1	33	2	8
Nasal voice	2	28	1	33	6	26
Vomiting	1	14	1	33	5	21
Blurring of vision	1	14	1	33	7	30
Irregular menses	1	14	—	—	1	4
Collapse of legs	—	—	—	—	1	4
Dyspnoea	2	28	—	—	—	—
Sleepiness	1	14	1	33	—	—
Fainting	1	14	—	—	—	—

impression is of diagnostic value for anyone who is acquainted with this sort of patient. Attention is immediately drawn by the short neck which was found in our material in 78% of the cases. This reduction of the cervical segment is indeed only apparent, since it is due to the basilar impression and only seldom to a Klippel-Feil deformity or even less frequently to a numerical reduction of cervical vertebrae.

Another very frequent finding is asymmetry of the skull or of the skull and face (68%). Garcin and Oeconomos (1953) give great importance to the anomalous positions of the head and particularly to limitations of head movement, painful or not. We have found in our patients a painful limitation of movement of the head in 53% and postural anomaly of the cephalic segment in 15% (Table VII). Other types of morphological and postural alterations were present in our patients. Table VIII shows that the more frequent findings were alterations of the deep reflexes in the sense of exaltation or only briskness in 74% of cases, associated with an abnormality of the plantar reflex (Babinski or indifferent reflex) in 65% of cases, and abolition of the abdominal reflexes in 59%.

Very often (65%) we find nystagmus (vertical, horizontal, or rotatory) and abolition of the gag (60%) and the palatal (53%) reflexes.

Disturbances of deep sensation (vibration and/or sense of position) were found in 59% of patients.

TABLE VII

ASSOCIATED MORPHOLOGICAL AND POSTURAL
ALTERATIONS

	(no.)	(%)
Morphological alterations of posterior fossa	27	40
Cranial-facial asymmetry	19	28
Brachycephaly	25	37
Brevis colli	52	78
Spinal deviations	18	27
Hammer-toes	2	3
Genu-valgum	3	4
Hypertelorism	5	7
Head tilting	10	15
Painful limitation of neck movements	35	53
Lumbar hyperlordosis	2	3
'Café-au-lait' spots	2	3
Microcephaly	1	1
Flat feet	6	9
Pes cavus	7	10
Pes varus equinus	2	3
Prognathism	1	1

Muscular weakness either in the four limbs or as paraparesis or hemiparesis was registered in 57% of patients. Cerebellar and vestibular disturbances appeared in 51% of our cases and Romberg's sign in 43%, spasticity in 46%, and hypotonia in 30%. There were some alterations of superficial sensation in 34% of cases, sometimes with atypical distribution but in others having the characteristics of syringomyelic dissociation. Other findings may be seen in Table's VIII and IX.

PURE ARNOLD-CHIARI DEFORMITY In the cases of isolated Arnold-Chiari deformity the dominant clinical picture is the result of cerebellar involvement characterized by motor inco-ordination and unsteadiness of gait or with lateral deviations, associated with vestibular signs and involvement of 9th and 10th cranial nerves. This syndrome was present in 100% of the cases.

PURE BASILAR IMPRESSION On the other hand in the cases of isolated basilar impression what is dominant in the clinical picture is the pyramidal syndrome in the form of deficit or release found in 100% of the cases, associated with disturbance of deep sensation which was also present in all patients. Involvement of other cranial nerves, particularly of the 5th (evidenced very often by hypaesthesia of one or both corneas) was also more frequently found in the patients with pure basilar impression.

We would like to emphasize the rarity of the syndrome of intracranial hypertension found in only 15% of our operated patients, all of them with basilar impression associated with Arnold-Chiari malformation, except one in whom there was no such malformation. Full details are shown in Table IX.

RADIOLOGICAL STUDIES

PLAIN RADIOGRAPH We have already mentioned the more important radiological features found in patients with basilar impression, drawing attention

to the several lines which confirm the diagnosis of the condition. Here we will only insist on some morphological aspects which, although not pathognomonic, are very frequently seen.

On the lateral views we may measure the basal-

TABLE VIII
GENERAL SYNOPTIC TABLE: SIGNS GROUP I-II

<i>Signs</i>	<i>Group I</i> (no.)	(%)	<i>Group II</i> (no.)	(%)	<i>Total</i> (no.)	(%)
Diminished visual acuity	4	12	3	9	7	10
Exophthalmos	—	—	2	6	2	3
Papilloedema or secondary optic atrophy	5	15	2	6	7	10
Involvement of II nd nerve	1	3	2	6	3	4
Involvement of V th nerve	18	54	16	48	34	51
Involvement of VI th nerve	5	15	5	15	10	115
Involvement of VII th nerve	5	15	9	27	14	21
Nystagmus	23	69	20	60	43	65
Diminished hearing	6	18	5	15	11	16
Paresis of soft palate	11	33	6	18	17	25
Abolition or diminished gag reflex	26	78	14	42	40	60
Abolition or diminished palatal reflex	20	60	15	45	35	53
Involvement of XI th nerve	9	27	9	27	18	27
Involvement of XII th nerve	6	18	7	21	13	19
Hypotonia	10	30	10	30	20	30
Hypertonia	15	45	16	48	31	46
Muscular weakness	20	60	18	54	38	57
Cerebello—vestibular disturbance	23	69	11	33	34	51
Hyperactive stretch reflexes	27	31	22	66	49	74
Decreased stretch reflexes	5	15	7	21	12	18
Hoffman reflex	7	21	7	21	14	21
Babinski reflex	24	72	19	57	43	65
Abolished or diminished abdominal reflex	21	63	18	54	39	59
Romberg's sign	22	66	7	21	29	43
Unsteady gait	22	66	12	36	34	51
Disturbance of superficial sensation	11	33	8	24	19	28
Syringomyelic disturbance of sensation	—	—	4	12	4	6
Disturbance of deep sensation	20	60	19	57	39	59

TABLE IX

COMPARATIVE TABLE OF THE CLINICAL SIGNS IN THE CASES B.I. OR A.C. ALONE AND B.I. + A.C.

<i>Signs</i>	<i>B.I. (7 cases)</i> (no.)	(%)	<i>A.C. (3 cases)</i> (no.)	(%)	<i>B.I. + A.C. (23 cases)</i> (no.)	(%)
Diminished visual acuity	1	14	—	—	3	13
Papilloedema or secondary optic atrophy	1	14	—	—	4	17
Involvement of V th nerve	4	57	—	—	14	60
Involvement of VI th nerve	1	14	—	—	4	17
Involvement of VII th nerve	1	14	—	—	4	17
Nystagmus	3	42	3	100	17	73
Diminished hearing	2	28	—	—	4	17
Paresis of soft palate	1	14	3	100	7	30
Abolition of gag reflex	4	57	3	100	19	82
Abolition of palatal reflex	4	57	3	100	13	56
Involvement of XI th nerve	2	28	—	—	7	30
Involvement of XII th nerve	—	—	1	33	3	13
Hypotonia	2	28	1	33	7	30
Hypertonia	5	71	1	33	9	39
Muscular weakness	6	85	1	33	13	56
Cerebellar and vestibular disturbances	4	57	3	100	16	69
Hyperactive stretch reflexes	7	100	2	66	18	79
Decreased stretch reflexes	1	14	1	33	3	13
Hoffmann reflex	2	28	—	—	5	21
Babinski reflex	7	100	1	33	16	69
Abolished or diminished abdominal reflexes	7	100	—	—	14	60
Romberg's sign	4	57	3	100	15	65
Unsteady gait	5	71	3	100	14	60
Disturbances of superficial sensation	4	57	1	33	6	26
Disturbances of deep sensation	7	100	1	33	12	52

sphenoidal angle of Schuller which may give evidence of an association of platybasia with a basilar impression.

Atlanto-axial dislocations may be seen and in some cases of intense basilar impression the posterior fossa takes the form of italic S.

On the anteroposterior views, vault asymmetries and petrous elevation may be seen.

In Towne's view petrous elevation with superior margins horizontalized are seen. The Hirtz view reveals anomalies of the foramen magnum, asymmetries of temporal fossae, and dislocation of the odontoid. The foramen magnum besides being asymmetrical very often presents the shape of a playing card—ace of hearts. Sometimes it is large, which would be suggestive of Arnold-Chiari malformation, in the opinion of Ricard and Girard (1949).

TOMOGRAPHY Tomography permits one to draw, quite easily, the Chamberlain and Fischgold lines and to confirm occipitalization of the atlas which could have been suspected on plain skull radiography.

Stereoscopic radiographs are particularly useful to examine the foramen magnum.

CONTRAST RADIOGRAPHS The procedure frequently used is lumbar myelography, the contrast material being carried to the level of the cranial-vertebral junction. In the cases of Arnold-Chiari malformation the positive contrast may take the shape of an open C (Garcin and Oeconomos, 1953; Malis, 1958, etc.). However, this appearance is not pathognomonic of the condition as has been demonstrated by Shapiro and Robinson (1955), who found similar shapes in two patients with high cervical meningiomas.

Pneumoencephalography may demonstrate absence of air entry into the ventricular system. Ventriculography may confirm the existence of an Arnold-Chiari malformation by showing an elongated 4th ventricle and cerebellar herniation in the spinal canal. Other authors prefer iodoventriculography, considering it as a better procedure than myelography in the diagnosis of the Arnold-Chiari malformation (Insausti and Matera, 1946; Verbiest, 1953; Canelas, Zaclis, Tenuto, and Cruz, 1956).

Vertebral angiography has been little used. In some cases it has been possible to see a downward displacement of the posterior inferior cerebellar artery into the superior part of the spinal canal, called the 'loop sign' by Spillane, Pallis, and Jones (1957).

We have sometimes seen this appearance during surgical operations. At present we rarely use techniques of contrast radiography because we

believe that it is possible to make the diagnosis of Arnold-Chiari malformation associated with basilar impression just by the analysis of the findings of the history and clinical examination. Furthermore, from the practical point of view, the treatment of these patients will not be changed by the existence or not of an associated Arnold-Chiari malformation. We reserve contrast radiological techniques for those cases in which there are possibilities of association with a tumour pathology.

CEREBROSPINAL FLUID

The information given by the spinal fluid examination is rarely helpful. The most frequent finding is an albumino-cytological dissociation. Occasionally one finds a manometric block, which is suggestive of the existence of an Arnold-Chiari malformation.

TREATMENT

Surgical treatment is the only one capable of giving appreciable results in the neurological complications of basilar impression and Arnold-Chiari malformation. Conservative treatment, by immobilization of the neck in a plaster collar and cervical traction, has been tried without convincing results (Phillips, 1955).

Indication for surgical treatment depends, however, on several factors. We do not advise surgery in cases of basilar impression without neurological symptoms, if the symptoms are slight or without progressive tendency. We prefer to keep these patients under observation, examining them periodically and sending them for surgery only if the clinical picture becomes worse. In some cases whose dominant symptomatology consists of nuchal pains we recommend the use of an orthopaedic collar.

Furthermore, the indication for surgery must be the clinical symptomatology and not the radiological degree of basilar impression. It is not rare to see cases of severe basilar impression in patients without neurological complaints or who present some symptoms unconnected with this condition and who ask for consultation for several other reasons. It is necessary, however, that these patients be followed up for a long time to permit finding the proportion in which slight symptomatology becomes worse or symptoms appear in asymptomatic cases.

Experience has taught us that the lesions in general seem to be progressive. Some symptoms, when they appear, no longer fade or only partially, even after surgical decompression, which should not be delayed until the clinical picture is severe. Suboccipital craniectomy, extended by a C1 and C2 laminectomy or a more extensive one, is used according to the case.

In order to achieve a good decompression we find

it necessary to open the dura mater and to cut all constrictive dural and arachnoid bands. Another very important detail is always to leave the dura open. In this we agree with the majority of authors who have studied the subject (Ray, 1942; Custis and Verbrugghen, 1944; Malis, Cohen, and Gross, 1951; Garcin and Oeconomos, 1953; Phillips, 1955).

The care to be given to the cerebellar tonsils when the Arnold-Chiari malformation is present has been a question of discussion by several authors. Some have made resections, but in general with disastrous results. We do not feel that the tonsils are a compressive agent after the dura has been opened and all bands sectioned.

In cases in which intracranial hypertension is present we explore the patency of the 4th ventricle and aqueduct which can be done quickly with a No. 7 Nelaton catheter, destroying any membrane or arachnoid process which by chance may be blocking the foramen of Magendie.

In cases without intracranial hypertension no intradural manipulation should be done and the arachnoid must be left intact (Malis *et al.*, 1951; de Barros, 1957, 1959). When there is intracranial hypertension we believe that a prior 3rd ventricle ventriculostomy may be necessary to control the hypertension. Some authors (Ricard, 1949; List, 1941; Ricard, 1953; Vidigal and Luccia, 1956; List, 1957) add to the decompression an occipito-spinal fusion by means of a bone graft, or immobilization of the neck with a plaster cast. We do not find these necessary.

RESULTS OF SURGERY

The surgical results have been satisfactory and support the conclusions of previous workers. We think that the cerebellar tonsils must be left intact in view of the disastrous results of resection obtained by others (McConnel and Parker, 1938).

In 32 cases operated on up to 1967, two (6%) were cured—that is, no complaints and normal neurological examination—19 (59%) remained much better than before operation, and four (12%) were unchanged.

The mortality (seven cases) of 21% was the same as that of Garcin and Oeconomos (1953). The best surgical results and the lowest mortality were in patients with Arnold-Chiari malformation in association with basilar impression (22 patients operated on with two cured, 14 improved, three unchanged, and three dead) (Table X). In seven cases with pure basilar impression there were four improved, one unchanged and two died. Of the three cases with pure Arnold-Chiari malformation, one is improved and two died.

TABLE X

OPERATIVE RESULTS

Types of malformation	Cases (no.)	Cured	Improved	Not improved	Deaths
B.I. + A.C.	22	2	14	3	3
B.I.	5	—	4	—	1
B.I. + Luxation of Atlas	2	—	—	1	1
	3	—	1	—	2
Total { no.	32	2	19	4	7
%		6	59	12	21

This is not in accordance with the literature, where it is stated that the Arnold-Chiari syndrome with or without basilar impression or other bony malformations gives better surgical results and lower mortality than cases without the Arnold-Chiari syndrome (List, 1941; Furtado, Marques, Oliveira, and Vicente, 1947; Gardner and Goodall, 1950; Scoville and Sherman, 1951; List, 1957). We believe that the poor results in patients with basilar impression alone is due above all to the neurological disorders resulting from the bony compression. In cases of the combined anomalies the cerebellar tonsils act as a cushion, protecting the medulla and spinal cord from the direct compression of the bony anomalies. We saw this in a patient who died from cardiac arrest, during operation, in whom the posterior lip of the occipital foramen, the arch of atlas and the axis were compressing strongly the medulla and the highest part of the cervical spinal cord, in such a way that the subarachnoid space was absent and the dura mater was so adherent to the neuraxis that attempts to separate it were unsuccessful.

In our series only two patients required re-operation. One of them required two further operations. The diagnosis of post-operative arachnoiditis was confirmed on both occasions. The other case with the same diagnosis had negative findings on re-exploration.

We would like to stress that these patients who had initial good results but who came back some time later with recurrence of symptoms, improved with re-exploration, including the one with negative re-exploration.

CONCLUSIONS

The overall findings and group differences are shown in Table X.

Our conclusions are based on Group I because we believe that they are of greater value as we have surgical and/or necropsy control in this group of patients.

Despite the small number of cases with basilar impression and Arnold-Chiari malformation, some conclusions are justified.

1. Basilar impression, associated with Arnold-Chiari malformation, is very frequent in the North-east of Brazil.

2. Race seems to be important, as, in 66 cases, only two were negroes, the other 64 whites or mulattoes.

3. The isolated forms of basilar impression and Arnold-Chiari malformation are relatively rare (seven and three cases respectively).

4. In patients with pure basilar impression, the clinical picture is basically a pyramidal syndrome of deficit or release associated with proprioceptive sensory disturbance.

5. In patients with isolated Arnold-Chiari malformation the clinical picture is represented by a cerebellar-vestibular syndrome, associated with 9th and 10th cranial nerve involvement.

6. The diagnosis can be made on the basis of plain radiograph; contrast radiographs being very often unnecessary.

7. The differential diagnosis between isolated forms of basilar impression and associated with the Arnold-Chiari malformation can be made in the majority of cases on a clinical basis.

8. Examination of the cerebrospinal fluid is not necessary, and sub-occipital puncture should be avoided.

9. The treatment is surgical and the operative results are, on the whole, satisfactory.

SUMMARY

The authors present a clinical study in which they analyse in detail the symptomatology of a series of 66 cases of basilar impression and Arnold-Chiari malformation, a relatively frequent condition in the North-east of Brazil.

Acquaintance with the clinical picture of the condition permitted diagnosis in almost every case on clinical evidence alone with plain radiograph of the skull, discarding contrast radiography.

It was possible to isolate two more or less characteristic clinical pictures in patients with radiographically demonstrated basilar impression. The first, pure basilar impression, is basically a pyramidal syndrome of deficit or release associated with disturbance of proprioceptive sensation and the second, combined basilar impression and Arnold-Chiari malformation, by a cerebellar-vestibular syndrome associated with cranial nerve involvement, particularly of the 9th and 10th nerves.

There is a discussion of indications for and results of surgical treatment by suboccipital craniectomy with a C1 and C2 laminectomy followed by opening of the dura, and section of all constrictive dural and arachnoid bands.

The dura must always be left open.

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