C R A N I O V E R T E B R A L A N O M A L I E S

A dissertation presented to the Department of Surgery, University of Nairobi, in part fulfillment for the Degree of Master of Medicine in Surgery (M.Med (Surg.).

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By

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DECLARATION

CANDIDATE:

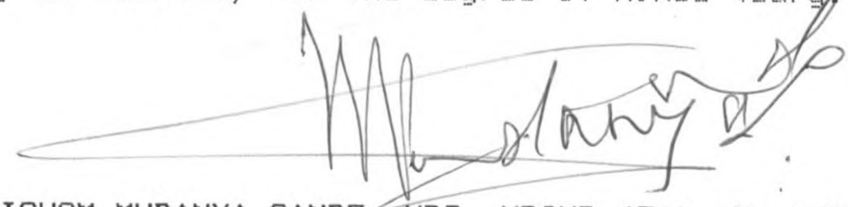
I certify that this is my original work done according to requirements and regulations of the University of Nairobi for the degree of M.Med (Surg.).

It has not been presented in any other University or elsewhere.

DR. GILBERT ADOGO MARANYA

SUPERVISOR:

I certify that this dissertation has been done under my supervision and according to the regulations and requirements of the University of Nairobi, for the degree of M.Med (Surg.).



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A C K N O W L E D G E M E N T

I am truly thankful to Professor G. M. SANDE for supervising me in this work. His constant encouragement and advice were invaluable.

I thank MWAI of the Medical Illustrations Department for the photography, my sister MIRIAM MARANYA for typing the work, and WILSON MAGETO for allowing me the use of his machine.

Lastly I give credit to my wife, MORAA ADOGO and son SAMORA ADOGO for domestic support.

C O N T E N T S

CRANIOVERTEBRAL ANOMALIES

MOTIVATION AND AIMS OF STUDY

Patients with developmental abnormalities at the craniovertebral junction caught my attention during training at Medical School and when working as a medical officer in Meru. The puzzling note was that, though the anomalies were developmental, they presented clinically with symptoms and signs in later life.

The patients I saw at the time were all from the ethnic groups of Kikuyu and Meru.

It was noted that more often than not the Medical Officers first seeing the patient did not make a correct diagnosis. This resulted in patients being subjected to unnecessary investigations, and causing a delay in diagnosis. Other patients were given diagnoses of conditions not treatable whereas craniovertebral anomalies treated appropriately and in good time have a good outlook.

The impetus was further strengthened by the desire to understand the anatomy and neurosurgical mystique of the craniovertebral junction.

The aims of this study were:

1. To understand the anatomy of the craniovertebral junction;

2. To define and classify craniovertebral anomalies and elucidate their pathological basis;

3. To look at the clinical and radiological features of these conditions;

4. To look into those factors that will result in early detection, early diagnosis, appropriate and timely treatment.

I N T R O D U C T I O N

Craniovertebral anomalies are developmental bony abnormalities at the craniocervical junction. They involve the occipital bone, the atlas and axis (1).

Knowledge of the anatomy of this region is essential to the understanding of these abnormalities. A brief study of the topographic and developmental anatomy of the occiput, atlas and axis bones will be undertaken.

ANATOMY OF THE CRANIOVERTEBRAL JUNCTION

TOPOGRAPHIC ANATOMY

This region is formed by the occipital bone, the atlas and the axis (2.3).

The occipital bone:

This bone has the following main parts: the basilar part, the squama and two condylar parts that articulate with the atlas (fig 1).

The bone has an external and internal surface. The external surface has lines for attachment of muscles. The internal surface has grooves for the superior sagittal sinus, the confluence of the sinuses and the transverse sinuses. The grooves mark off two cerebral fossae above and two cerebellar fossae below. The falxe cerebri, the tentorium cerebelli and the small falxe cerebelli are attached to the edges of the grooves.

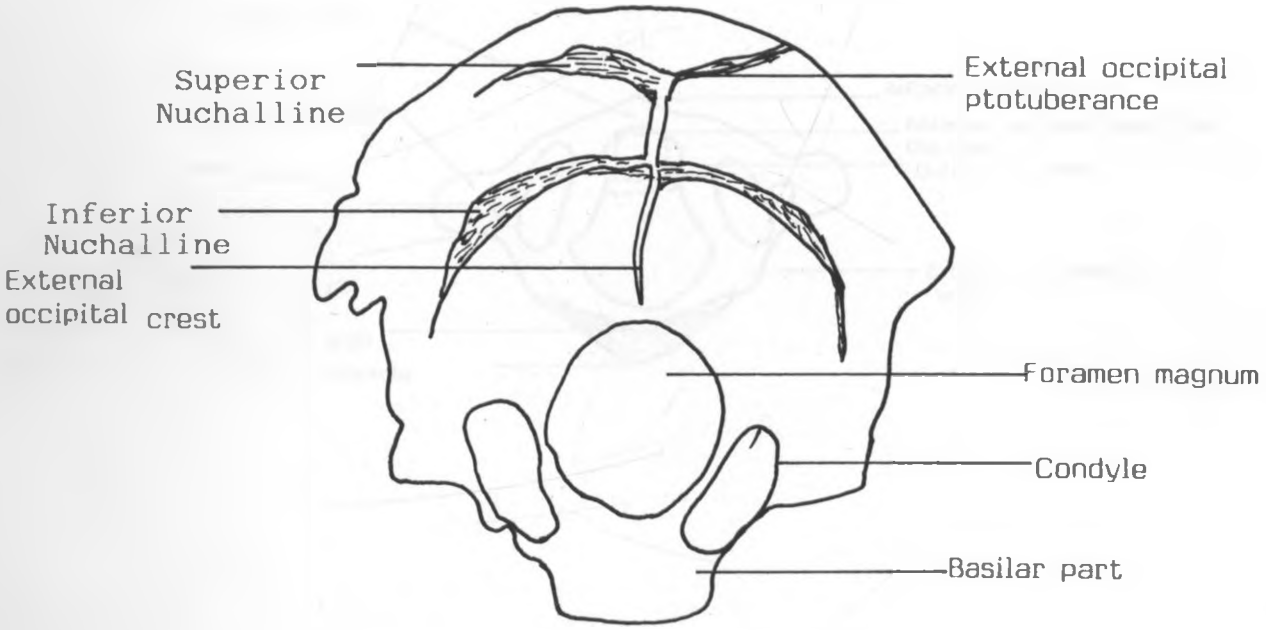
The bassilar part of the bone forms a groove that lodges the medulla with its coverings and the vertebral arteries.

The atlas:

The atlas (first cervical vertebra) consists of slender arterior and posterior arches united on each side by a lateral mass on which are situated the articular facets and the transverse process (fig 2).

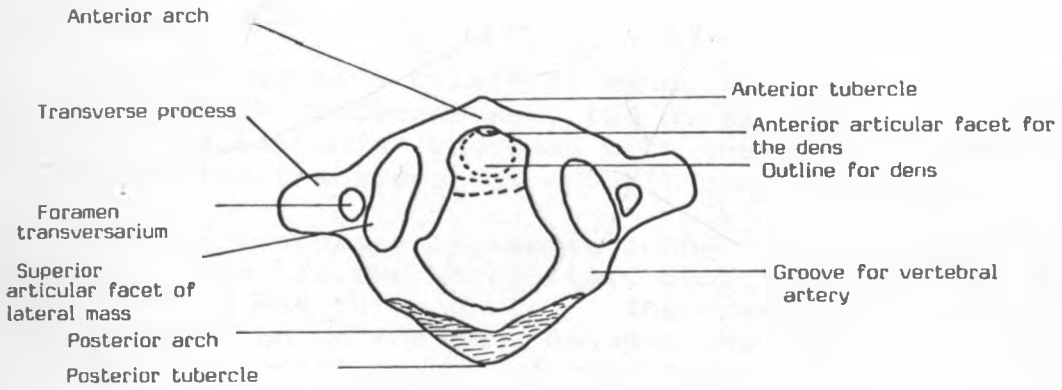
Most of the body (centrum) of the atlas joins the axis to form the dens (odontoid process). The facets articulate with the occiput above and the axis below.

The vertebral artery grooves the superior surface of the posterior arch where it joins the lateral mass, and curves medially after emerging from the foramen transversarium. The anterior arch has a facet on the middle of its posterior surface for articulation with the dens of the axis.



Occipital bone (Inferior Aspect)

Fig. 1



The atlas (superior Aspect)

Fig. 2

The axis:

The axis (second cervical vertebra) is the strongest cervical vertebra but with the smallest transverse process (fig 3). Its superior artricular facets transmit the weight of the head to its body, leaving the dens free to act as a pivot.

The atlanto-occipital joints::

The skull is joined to the atlas by a pair of atlanto-occipital synovial joints between the occipital condyles and the superior articular facets of the atlas, and by the anterior and posterior atlanto-occipital membranes, which connect the arches of the atlas with the margins of the foramen magnum (fig 4).

Movement at the atlanto-occipital joints takes place in two principal directions; nodding or bending the head forwards and backwards and lateral flexion of the head on the neck.

The atlanto-axial joints:

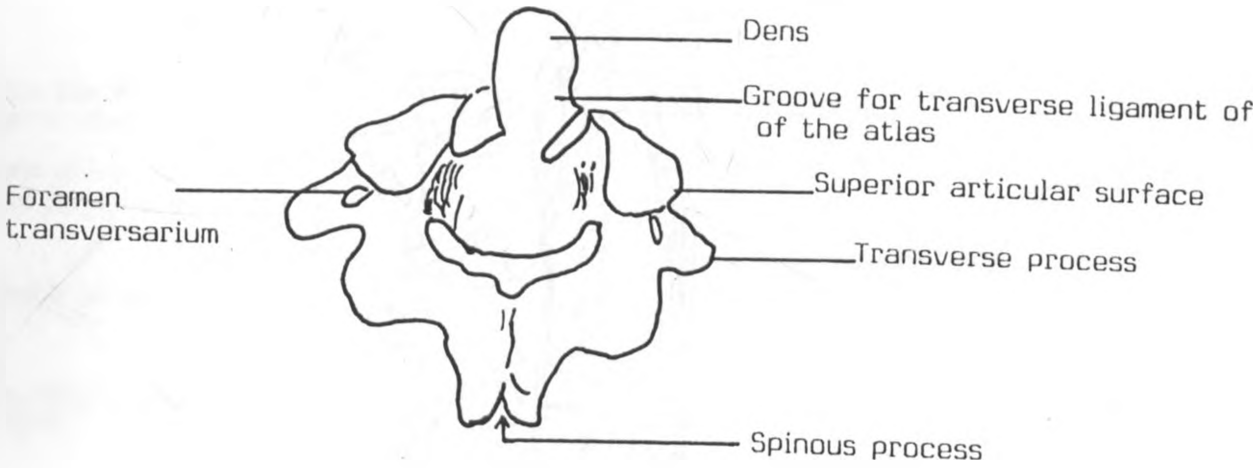
The atlas and the axis have bilateral synovial joints between their opposed articular processes and, two median synovial joints formed by the articulation of the dens with the arterior arch of the atlas and with the transverse atlantal ligament (fig 5).

In addition various accessory ligaments connect the axis to the atlas, and the dens to the occipital bone. The cruciform ligament of the atlas has three parts. The transverse ligament of the atlas passes behind the dens between the tubercles on the medial sides of the lateral masses of the atlas. From the middle of the transverse ligament a superior longitudinal band of the cruciform ligament passes upwards to the arterior edge of the foramen magnum, and an inferior longitudinal band of the cruciform ligament passes downwards to the back of the body of the axis.

The occipito-axial ligaments:

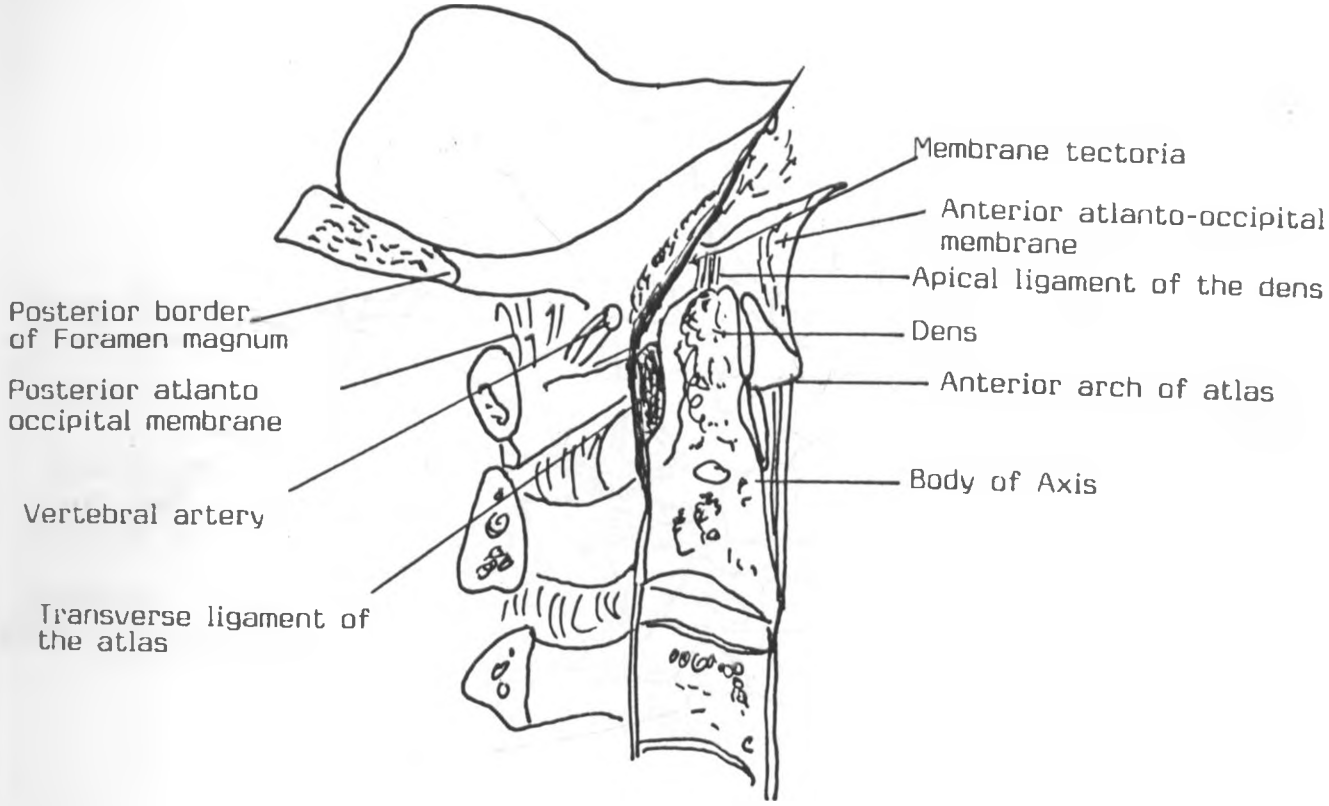
The axis is connected with the occipital bone indirectly by the longitudinal bands of the cruciform ligament, and more directly by the apical and alar ligaments of the dens and by the membrane tectoria (fig 4 and 5).

The combined movements at the three atlanto-axial joints serve to effect rotation of the head around a vertical axis, the skull and atlas moving as one around the dens. Excessive rotation is checked by the alar ligaments and to a lesser extent by the accessory atlanto-axial ligaments.



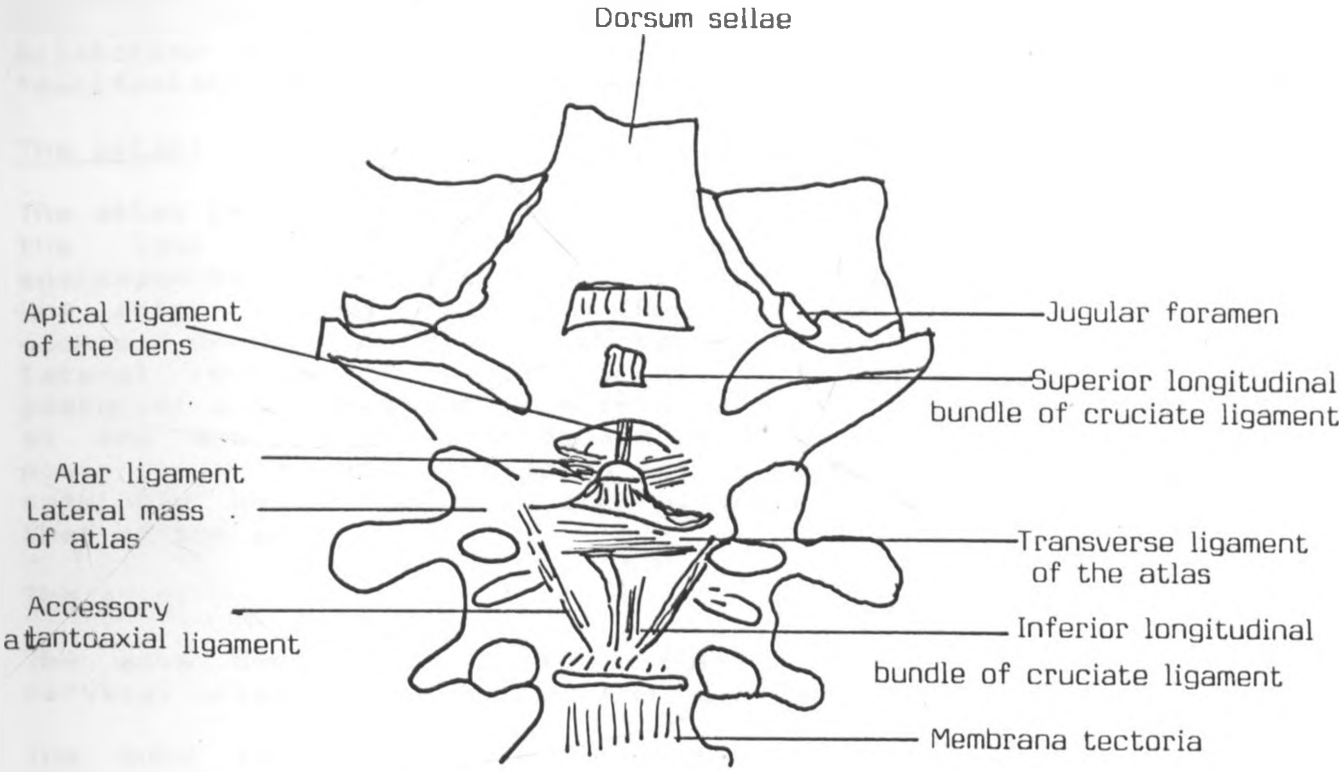
The axis vertebra (From behind and above)

Fig. 3



Amedian sagittal section through the occipital bone and the first to third cervical vertebra

Fig. 4



Hindview of base of skull showing the main ligaments that connect the occipital bone, the atlas and axis.

Fig. 5

DEVELOPMENTAL ANATOMY OF THE CRANIOVERTEBRAL JUNCTION

The normal osseous development (1,28) is described in fig [6]

The occiput:

The posterior part of the skull is formed by coalescence of four or five occipital sclerotomes which correspond to the primary roots of the hypoglossal nerve [4]. The most caudal of these sclerotomes later loses its identity when it becomes united with the proximal parts. Failure of fusion of this last occipital scletotome with the remaining occipital segments is termed "manifestation of the occipital vertebra".

The atlas:

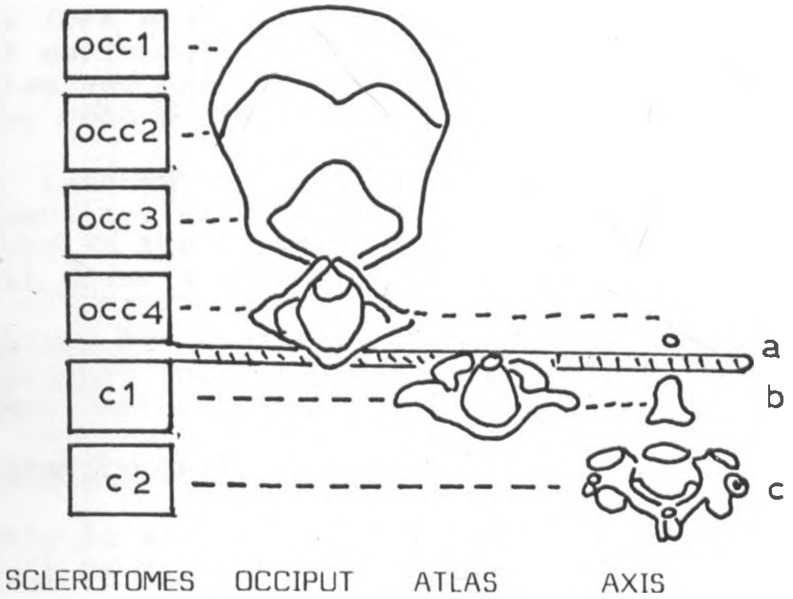
The atlas is formed by three elements, namely, the caudal half of the last occipital sclerotome and the cranial and caudal semisegments of the first cervical sclerostome [1]. The body of the atlas is lost early; it furnishes the main mass for the odontoid process. The atlas contains one anterior median and two lateral centres of ossification. The median cleft in the posterior arch resulting from this development is normally closed at the end of the first decade of life. Non-fusion of the posterior arch results in spina bifida posterior. Occasionally a seemingly absent poserior arch of the atlas may be fused with that of the axis.

The axis:

The axis develops from three primitive vertebrae. The second cervical sclerotome furnishes the main mass.

The dens is formed by two centres of ossification; the major distal portion represents the original body of the atlas, whereas the tip, the 'ossiculum terminale' is derived from the body of the proatlas (the last occipital sclerotome).

Anomalous development of the second and third cervical vertebral bodies and of other cervical vertebrae is not rare [1].



Schematic drawing to show ontogenetic development of the occipital bone, atlas and axis. The hatched bar represents the craniovertebral boundary. The occipital squama and proatlas are drawn as viewed from below, and the atlas and axis as viewed from above. In this drawing a indicates ossiculum terminale of odontoid process, derived ng from proatlas, b odontoid process, representing original body of atlas, and c , axis.

Fig. 6

CRANIOVERTEBRAL ANOMALIES AND THEIR PATHOLOGICAL BASIS

Congenital malformations of the occipitocervical area were previously considered as mere anatomic curiosities and were of interest first to the anatomists and anthropologists and then to radiologists. Only recently have they emerged into the more practical field of clinical neurology.

The principal craniovertebral anomalies [1,5] are:

1. OCCIPITALISATION OF THE ATLAS

2. BASILAR INVAGINATION

3. CHRONIC ATLANTO - AXIAL DISLOCATION

Some cases form a miscellaneous group impossible to classify [5]. A range of malformations is frequently associated with anomalies of the atlas and axis, some of which may be quite atypical. The latter have been grouped as sub-occipital dysplasia [4].

The most interesting feature of craniovertebral anomalies is their diversity: each type may vary in degree, in its clinical effects and in the pattern of its association with other neural or skeletal defects in the neighbourhood. [6]

The anomalies per se have little pathological significance and usually produce no clinical symptoms except when there is encroachment on the neural canal. [1]

OCCIPITALISATION OF THE ATLAS

This anomaly is also called assimilation of the atlas to the base of the skull or atlanto-occipital fusion.

The essential point in the diagnosis is some degree of bony union between the skull and the atlas [1,5,7]. Such synostosis may affect the anterior arch of the atlas fusing it to the lower end of the clivus, or the posterior arch, which is then fused tot he squamous occiput just below the posterior rim of the foramen magnum. Both anterior and posterior fusions may be present in the same patient. The lateral masses may be occipitalised on one or both sides or these may be synososes at one or both atlanto-occipital joints.

Occipitalisation of the atlas was first mentioned in the literature by R. COLUMBUS (1752) [8], and has been much discussed in the anatomic literature of the last two centuries [1].

In general, atlas assimilation is not an isolated malformation, but is found associated with basilar impression or with fused cervical vertebrae [4].

There are several possible causes responsible for bony union of the atlas with the occiput. [1]

1. In a small group of cases fusion is the result of a pathological process (such as arthritis or tuberculosis) occurring in foetal or in early postnatal life.

2. Some authors believe that intrauterine trauma, namely, pressure on the hyperextended foetal head is an important factor in the development of the deformity.

3. In the majority of cases however, the condition occurs on a purely developmental basis and represents a variation in the craniovertebral boundary.

   The assimilated atlas frequently shows maldevelopment, spina bifida or even absence of its posterior arch.

In atlas assimilation neurological manifestations occurs in four conditions [4].

1. When the odontoid process is excessively mobile, due to the absence of the transverse ligament, leading to compression of the medulla and spinal medulla by dorsal movements [9].

2. Due to compression of the vertebral arteries, especially by head movements, if the arteries do not find an adequate entry through the base of the skull.

3. Due to narrowing of the foramen magnum by the assimilation itself.

   The odontoid process comes in contact with the medulla oblangata because the anterior arch of the assimilated atlas is not only high but also posterior in position as a result the odontoid process often lies partly or wholly within the foramen magnum.

4. Due to the regularly found high placement of the odontoid process [7].

Pressure from behind comes from the posterior arch of the atlas, the posterior lip of the foramen magnum or from a thick band of dura at the junction of the cranial and spinal dura.

Occipitalisation of the atlas is frequently associated with other anomalies of the craniovertebral junction. Particularly frequent is fusion of cervical vertebrae [6]. Others are basilar invagination and atlanto-axial dislocation.

Tonsillar herniation and syringomyelia are frequently associated with atlas assimilation. Damage of the more rostral cranial nerves is infrequent. [4].

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Occipitalisation of the atlas can usually be diagnosed by good plain films. For occipitalisation of the anterior or posterior arches good lateral films in extension and flexion will show the abnormality although tomograms may be needed for confirmation.

Plain films taken through the open mouth and centered on the odontoid may suggest the presence of synotosis at the atlanto-occipital articulations but anteroposterior tomograms are required for confirmation of such fusion, and in order to establish a diagnosis of occipitalisation of the lateral masses [7,10].

All patients showing an effective anteroposterior diameter of the foramen magnum of less than 19mm (odontoid to either posterior arch of the atlas or posterior lip of the foramen magnum) have symptoms or signs of disease in the lower medulla or upper spinal cord.

When the diameter is more than 19mm and symptoms and signs of spinal cord compression are present, this may be due to dural bands [10].

PLATYBASIA AND BASILAR INVAGINATION

PLATYBASIA denotes an increase in the breadth (obtuseness) of the basal angle of the skull which is the angle made by the intersection of the plane of the spheroid with the plane of the clivus [6].

Platybasia is determined by measuring the basal (Boogard's) angle [11]. In a lateralfilm of the skull, the plane of the spheroid (plane of the anterior fossa) is determined by drawing a line from the nasion (nasofrontal suture) to the tuberculum sellae [5], centre of the pituitary fossa [12], or posterior clinoids [1]. The plane of the clivus is determined by drawing a line from the tuberculum sellae, centre of the pituitary fossa or posterior clinoids to the anterior lip of the foramen magnum. Although the point of intersection of the two lines varies, the resulting variation in the basal angle does not exceed a few degrees.

Boogard [11] on direct measurement found the basal angle to vary between 103.5 degrees and 131.5 degrees. List [1] considered the normal range of the angle to be 115 - 140 degrees. McRae and Barnum [7], who have an extensive experience in the radiological analysis of craniovertebral anomalies, consider platybasia present when the basal angle equals or exceeds 145 degrees. Platybasia per se has no clinical significance.

BASILAR INVAGINATION (BASILAR IMPRESSION) is a deformity of the base of the skull consisting of an elevation into the cranial cavity of a variable part of the bony rim of the foramen magnum [1,5,6].

Basilar invagination ('impressio baseos cranii') of Berg and Retzinus [13], and first diagnosed radiologically in life by Schuller [14], aroused interest only after Chamberlain [15] simplified its radiological diagnosis and stressed its frequency.

Two forms of basilar invagination are differentiated according to their genesis: primary and secondary basilar invagination.

Primary basilar invagination

The form this study addresses, represents a congenital bone malformation involving the posterior cranial fossa. Its cause is not known and genetic factors are assumed to be of primary importance [4,16].

Secondary basilar invagination

This is always associated with a disease which leads to a fundamental structural change of the bone. As the base of the skull softens, the weight of the cranium produces an indentation of the base on the upper cervical spine. In early childhood the indentation may also arise from an expansion of the soft bones of the posterior cranial fossa caused by the excessive but physiological growth of the cerebellum [4]. Secondary basilar invagination is relatively rare.

Causes of Secondary Basilar Invagination

1. Paget's disease: Most frequently described cause of secondary basilar invagination [17,18]. The neurological symptoms which may be found in this condition are often due to the narrowing of the spinal canal, as well as the concomitant secondary basilar impression.

2. Osteomalacia, usually caused by malnutrition [19,20]. Disordered bone metabolism may also result from coeliac disease and after gastric surgery [21].

3. Hypoparathyroidism [22]

4. Hyperparathyroidism [23,24] Interestingly the association with basilar invagination is associated with mild rather than severe forms of the disease.

5. Fibrous dysplasia

7. Gargoylism (Pfaundler - Hurler syndrome)

8. Chondrodystrophy (achondroplasia)

9. Trauma May result from severe injuries resulting in endocranial displacement of the occipital bone. Extremely rare as patients with such injuries rarely survive.

Pathogenesis of clinical manifestations in primary basilar invagination

In explaining the pathogenesis of the clinical manifestations the following points should be borne in mind [4]:

1. X-Ray and pathoanatomical investigations reveal only the degree of narrowing about the foramen magnum. The bony space restriction may be exacerbated by an accompanying cerebellar tonsillar herniation.

2. Post-mortem and operative observations often reveal adhesions of the arachnoid.

3. Pathological, anatomical and angiographic findings and certain clinical features prove that there may be circulatory disturbance along the vertebrobasilar axis.

4. Despite the fact that the bone malformation is clearly congenital, the clinical manifestations usually do not progress before middle age.

Foramen magnum narrowing can result in pressure damage of the medullary structures of the brain, nerves and vessels. This direct mechanical effect has been emphasized by some [5].

The fact that the dens is higher than usual produces a kyphotic bending of the brain stem. The medulla oblangata and the upper spinal medulla may be pressed [23], resulting in pressure necrosis and demyelination. The pyramidal tracts are especially affected.

Adhesions of the arachnoid are not always found. Thismeans that pressure alone is of cardinal importance for the development of the clinical picture.

There may be primary or secondary cerebellar tonsillar herniation, which may enhance the pressure effect.

Very often post-mortem or operative observations show extensive arachnoid adhesions in the region of the foramen magnum associated with fixing of the frequently extended cerebellar tonsils or a glueing together of the cranial nerves [25].

Arachnoiditis in craniocervical malformation may not be simply the result of inflammatory or haemorrhagic processes. It may best be explained by a number of microtrauma, occurring with head movements. Physiological or pulse movements of the brain may also play a part, as the cerebellar tonsils press into the foramen magnum and rub rhythmically against the surrounding structures. These injuries are exercabated when the spinal medulla and medulla oblangata are fixed by the arachnoid adhesions.

Vascular changes which are due to bone pressure and arachnoiditis are important in the development of neurological symptoms.

The losses in the visual field in patients with basilar impression are often uncharacteristic [26]. They are probably explained by vascular and arachnoid changes

The vascular changes, arachnoid adhesions and the frequent cerebellar tonsillar herniation are sufficient explanation for the rather meagre X-Ray changes in the craniocervical malformations, compared to the neurological findings. On the other hand it is unclear why impressive X-Ray appearances of bone change may remain asymptomatic.

Measurement of basilar invagination

The rim of the foramen magnum lies at the lowermost part of the posterior cranial fossa. The assessment of whether a given patient has or does not have basilar invagination is not always straightforward [6].

The following are the classical methods used in the diagnosis of this anomaly.

1. Boogard's (Basal) angle [11]

This has already been described. It is not much used nowadays. It used to be considered that higher values >145 degrees were characteristic of basilar invagination. Average range is 115 - 145 degrees.

2. Chamberlain's line [15]

This is a line drawn on the lateral radiograph of the skull from the "dorsal lip of the foramen magnum to the dorsal argin of the hard palate". Chamberlain considered that projection of the odontoid process above this line constituted basilar impression.

Various workers have measured the position of the odontoid with respect to this line and there is quite a variation. For instance:

Poppel [17] in a study of 102 radiographs found the average position of the odontoid to be 0.6 +/- 3.3mm. Bull [16] in a study of 120 radiographs found the average position of the odontoid to be -2.86 +/- 3mm.

Minor drawbacks to the use of Chamberlain's line are [6]:

a) The posterior lip of the foramen magnum, one of the points of reference, is often difficult to define in the average lateral radiograph.

b) That in certain types of basilar invagination the posterior lip of the foramen magnum is itself invaginated.

3. McGregor's Line [12]

McGregor substituted for Chamberlain's line, a line drawn "from the upper surface of posterior edge of the hard palate to the most caudal point of the occipital curve in the true lateral X-Ray". Advantage over Chamberlain's line is that the posterior point is both fixed and easy to define.

Position of the odontoid by McGregor on measurement of 203 Bantu skulls was -1.32mm +/- 2.62mm. Bull's [16] average was -0.39 +/- 3.02mm.

4. Bull's angle [16]

This is determined by the plane of the hard palate and the plane of the atlas vertebra. Patients with angles of 13 degrees or more should be considered as having basilar impression.

5. Fischgold's line [27]

In the anterposterior film of the skull (or better still in the transoral projection), a line joining the lower extremities of the mastoid processes passes through the atlanto-occipital articulations and the tip of the odontoid is often situated on the same line. In cases of basilar impression, one or both atlatno-occipital articulations are elevated above this line.

6. The digastric line [27]

In view of the variability in the size of the mastoid processes Fischgold and Metzer later proposed a more constant baseline, drawn between the two digastric grooves (each groove is situated at the junction of the medial aspect of the mastoid process with the base of the skull). This digastric line can often be drawn on a good transoral film and is nearly always easy to define on an anteroposterior tomogram. The digastric line usually passes well above the occipital condyles and the odontoid.

7. Tomography of the skull

Tomograms are of great value in the diagnosis of basilar impression [5].

8. Computerised tomography scan [24]

This is a new radiodiagnostic method and is the most accurate in diagnosis of basilar invagination.

CHRONIC ATLANTO-AXIAL DISLOCATION

In most of these cases the type of dislocation is an anterior dislocation of the atlas on the axis. Posterior dislocations have been reported and these can only occur if the odontoid is separate [28].

The essential sign of atlanto-axial dislocation is widening of the joint space between the anterior arch of the atlas and odontoid (A - O interval).

The normal atlanto-odontoid interval is not greater than 2.5mm in adults [29], and normally there is no movement of the atlas on the axis with flexion of the head. However in children there is movement of the atlas on the axis with flexion and the A - O interval may be as large as 4.5mm in this position.

Measurement of the A - O interval is appropriate for detecting an atlanto-axial dislocation as long as the odontoid process is intact and is attached to the body of the axis. If the structure of the odontoid is altered by trauma, infection, tumour, or by a congenital defect, a dislocation can occur with the odontoid riding forward with the anterior arch of the atlas due to the intact transverse atlantal ligament, and the A - O interval will not be altered. [28]

In such cases it is more important to measure the smallest anteroposterior diameter of the canal. In the anterior dislocations this measurement will be from the posterior arch of the atlas to the nearest point on the posterior surface of the axis-dens complex. In the less frequent posterior dislocations this measurement will be from the posterior arch of the atlas to the posterior surface of the dens (or the anterior arch of the atlas if the dens is absent).

Some degree of lateral dislocation often accompanies an antero-posterior dislocation.

Giacomini [30] described the first case of congenital atlanto-axial dislocation.

Atlanto-axial dislocation may be congenital, but more frequently is acquired, due to trauma, infection, and rheumatoid arthritis of atlanto-axial joint.

All cases of anteroposterior atlanto-axial dislocation, regardless of aetiology, can be classified as: [28, 311]

I Those caused by 'incompetence of the odontoid process;

II Those caused by 'incompetence of the transverse atlantal ligament';

III Maldevelopment of the lateral articulating facets between the atlas and the axis.

This is summarised in the table below:

TABLE 1: CLASSIFICATION OF ATLANTO-AXIAL DISLOCATIONS

I Incompetence of the odontoid process

A) **Congenital**

1) Type I: Separate odontoid - "os odontoideum"
2) Type II: Free 'apical' segment - "ossiculum terminade"
3) Type III: Agenesis of odontoid base
4) Type IV: Agenesis of apical segment
5) Type V: Agenesis of odontoid process

B) **Traumatic**

1) acute
2) chronic

C) **Infections**

D) **Tumour**

1) Primary
2) Secondary

II Incompetence of the Transverse Atlantal Ligament

A) **Congenital**

1) Idiopathic
2) Mongolism

B) **Traumatic**

1) Acute
2) Chronic

C) **Hyperaemic**

1) Infections
   a) viral
   b) bacterial
   c) granulomatous
2) Rheumatoid
   a) arthritis
   b) ankylosing spondylitis

III Maldevelopment of the lateral articulation facets between the atlas and the axis

The congenital malformations of the odontoid may best be appreciated diagramatically (fig 7).

The pathophysiology of the dislocation associated with a fractured odontoid is identical with that of a congenitally separated odontoid. Once a fracture occurs, depending upon the

Agenesis of the odontoid

Agenesis of odontoid base

Normal axis

Agenesis of apical segment

"Ossiculum terminale"

"Os odontoideum"

Sites of dysgenesis in the development of the odontoid

Fig. 7

severity of dislocation, it can cause symptoms of cord compression immediately, or by producing an unstable articulation, cause repeated small episodes of cord trauma over many years leading to a 'delayed myelopathy'. [32]

Because of the similarity in clinical course it is often difficult to distinguish between a cause of fractured odontoid and a congenitally separate odontoid even if a history of injury is available.

The table below (table 2) compares the various distinguishing features of the two entities.

Table 2: **Differentiating features of congenital vs traumatic separation of odontoid**

	Congenitally separate odontoid	Fractured odontoid
History of trauma	Often present	Always present but may be remote
Site of separation	Usually between base and apical segment of dens (above superior articular facet)	Usually between base of dens and body of axis (below superior articular facet)
Line of separation	Always smooth and corticated	Acutely irregular and not corticated
Associate congenital anomalies	Often present	Absent

In considering congenital incompetence of the transverse atlantal ligament, the idiopathic group, with no known cause, the diagnosis is one of exclusion. Section of the odontoid process by a shortened transverse ligament has been reported [33].

In mongolism incompetence of the transverse atlantal ligament is seen in 20 - 30% of cases [28]. It is thought that the dislocation may be a reflexion of the generalised, looseness of joints seen clinically in this condition.

In traumatic incompetence of the transverse atlantal ligament, acute trauma may occasionally rupture the ligament with the dens remaining intact. Included in this category are the dislocations that are due to increased stress on the transverse atlantal ligament from a congenital anomaly of the upper cervical spine.

ASSOCIATED BONY ANOMALIES OF THE CRANIOVERTEBRAL JUNCTION

Klippel-Feil Syndrome

This includes complete fusion of the cervical vertebrae, reduction in the number of cervical vertebrae and partial or complete coalescence of two or more cervical segments.

When neurological complications do arise in this syndrome, they are not directly due to the fused cervical vertebrae but to associated bony or neural malformations at the craniovertebral junction. [6]

Spina bifida posterior or spondyloschisis of the atlas.

Asymmetry of the lateral facets

Other stenotic lesions [21] that may narrow the neural tube are chondrodystrophy (achondroplasia), occipital dysplasia in which the foramen magnum is small and deformed as a result of premature fusion of one or more occipital synchodroses, and congenital spinal stenosis.

The acquired condition of chronic fluoride intoxication may stenose the neural canal. This results from thickening of the skull base, ligamentous calcification and protruding osteophytes that may compress the medulla and spinal cord, producing a severe myelopathy.

ASSOCIATED ANOMALIES OF THE NEURAXIS

Arnold-Chiari malformation

This term encompasses a series of abnormalities consisting of two major components: (1) a tongue of cerebellar tissue which extends posteriorly to the medulla and spinal cord for several centimeters into the cervical canal; and (2) a displacement of the medulla into the cervical canal, along with the inferior part of the 4th ventricle.

Since the malformation is already existent at birth whereas the clinical expressions of it appear later, the operation of secondary factors seems important, namely:

Progressive effects of hydrocephalus, hydromyelia and syringo;yelia, progressive cerebellar herniation and atrophy, and progressive traction and stretch injury of the spinal cord [4].

Syringomyelia and syringobulbia:

Two forms are distinguished [4]: a 'non-communicating' form associate with intramedullary tumours, traumatic paraplegia and some degenerative conditions, and 'communicating syringomyelia' which may be developmentally determined and results from a persistent dilatation of the central canal under pressure with subsequent rupture of fluid into the surrounding neural tissue.

Syringomyelia is commonly associated with craniovertebral anomalies [5, 16]. The association may be considered thus [6]:

i) Skeletal and neural defects are not directly related. Majority of cases are probably of this type; both defects being manifestations of disordered development.

ii) Acquired intramedullary cavitation. This may result from ischaemic myelomalacia or obstructive hydromyelia.

CLINICAL FEATURES

Neurological symptoms may logically be grouped [4] into:

1 Bulbopontocerebellar disturbances

Most common is nystagmous. Others include ataxia. Dysphagia and speech disorders are part of cranial nerve disturbances and bulbar, suprabulbar and cerebellar disturbances. Respiratory disturbance is rare.

2 Long tract disturbances

Pyramidal tracts are most commonly affected. Posterior tract symptoms occur to varying degrees.

3 Cranial nerve symptoms

The accessory nerve is often involved but the disturbances are mild.

Others are hypoglossal paralysis and vagus (cord paralysis), Glossopharyngeal, auditory, facial and trigeminal nerves are rarely involved.

4 Disturbances of the anterir horn motor nerves

This presets with various degrees of flaccid paralysis and atrophy of the muscles of the neck, shoulder, arm and hand.

5 Cerebrospinal fluid circulation disorders

These will be symptoms of increased intracranial pressure. Trauma of varying severity is often the precipitating factor [1,7,23,32,36]. Long-term pressure on the cervical spine (e.g. carrying of loads on the head as in India [36] and Africa) seems to play a part. This is probably due to a cumulative effect of microtrauma.

The triad of **signs** [34] which characterise craniovertebral anomalies are:

1 an abnormal neck;
2 cerebellar signs;
3 spastic quadriparasis

An abnormal neck includes the following:

i) Short neck with a low hairline,

ii) 'Hunched' appearance, in which the head and neck are held as if pushed forwards on the shoulders,

iii) Limited neck movement with or without pain.

Cerebellar signs include ataxic legs, ataxic arms and nystagmus.

Spastic quadriparesis is upper motor neurone signs in the legs and the arms.

RADIOLOGICAL INVESTIGATIONS

These are the mainstay of diagnosis in craniovertebral anomalies. These may be grouped as:

I PLAIN X-RAYS AND TOMOGRAPHY of the head and neck.

II COMPUTERISED TOMOGRAPHY:

This is a new technological advance and is the most accurate so far.

III CONTRAST MEDIUM STUDIES

Tailored to the individual patient, contrast techniques should always be employed even in patients in whom the bone malformations seem to account for the neurological symptoms. In this way missing a coincidental space occupying lesions in the posterior fossa and neural malformations underlying the skeletal defect is avoided. [4, 6]

Angiography is emphasised over contrast studies of the cerebrospinal fluid for the reason that vascular factors are more important in the genesis of symptomatology in these anomalies.

A **Vascular studies**

Vertebral angiography is the most important method of investigation. Carotid angiography only provides complementary information [4].

1 Vertebral angiography

a) The angiographic findings associated with bone abnormalities of the craniovertebral junction are:

i) Vascular variants and anomalies
ii) Abnormal vascular pathways
iii) Obstructive vascular changes which may be permanent or motion dependent.

b) Angiographic findings in associated malformations of neural structures

In general a lower loop of the posterior inferior cerebellar artery is seen to extend into the upper cervical spinal canal (loop sign [6]) in a case of marked bone abnormality at the craniovertebral junction corresponds to a low position of the cerebellar tonsils.

Other than for the 'loop sign', cerebellotonsillar herniation may be demonstrated by increased vascularity partly due to the abnormally low position of the choroid plexus of the fourth ventricle and partly to vascularisation of the meninges.

2 Carotid angiography

In combination with vertebral angiography it permits survey of the entire intracranial circulation. Particularly where vertebral angiography shows a reduced supply to the posterior fossa, determination of an adequate collateral articulation from the carotid system is important.

Approximation of the size of the lateral ventricles may be made by the size of the arch of the anterior cerebral and pericallosal arteries and by the course of the thalamostriate and subependymal veins.

B CONTRAST STUDIES OF THE CEREBOSPINAL FLUID SPACES

A number of associated malformations can be definitely diagnosed by visualisation of the CSF spaces with the help of contrast media.

The myeloencephalagraphic techniques use either positive or negative contrast.

1. Pneumoencephalography and air myelography

2. Myelography with positive contrast

3. Ventriculography

T R E A T M E N T

Conservative procedures e.g. head immobilisation using a collar or sling may provide temporary relief.

Surgical treatment is advised for all patients with neurological signs [37].

For basilar invagination and occipitalisation of the atlas surgical treatment consists of suboccipital craniectomy and laminectomies of the upper cervical vertebrae. If indicated, the arachnoid adhesions are cleared as far as possible. [14]

Surgical treatment for congenital atlantoaxial dislocation depends on whether the dislocations is reducible or not.

In reducible dislocation due to congenital incompetence of the odontoid process posterior fusion [40,45,46] is recommended following reduction with extension of the head or by tongs.

Reducible dislocation due to congenital incompetence of the transverse atlantal ligament is a potentially serious disorder as the site of compression is higher with intact odontoids - being either high cervical or medullary [41]. After reduction, all should have posterior fusion.

As for non-reducible dislocations, the morbidity and mortality is high [1,36,41,42]. However operative intervention is imperative.

Unlike reducible dislocation where stabilisation is the primary goal, the treatment of non-reducible dislocations aims at decompression and stabilisation to prevent recurrent difficulties. [2]

Posterior decompressive procedures have a high risk [41] because of the necessity to flex the head to achieve adequate exposure.

An anterior approach to the formen magnum and atlanto-axial region either through a transoral [28] or transcervical route [43,44] offers the advantage of [47] performing the operation with the head in extension rather than flexion. The only drawback however is that a second operation for stabilisation is necessary.

Suboccipital craniectomy and laminectomy of upper cervical spinal without stabilisation is catastrophic in atlanto-axial dislocations as the instability is increased [47]. In fact, laminectomy is contraindicated, the procedure of choice being atlanto-axial or occipitocervical fusion after reduction of the dislocation.

Complications of operation include death which may be due to haematomyelia following decompression [41], respiratory paralysis due to oedema or extraneous pressure on the medulla and spinal cord, and hydrocephalus.

PATIENTS AND METHODS

A study of 27 patients with craniovertebral anomalies was done both retrospectively and prospectively spanning the period 1981 - 1987. The retrospective study involved 25 patients and the prospective 2 patients.

The files of these patients were obtained from the Records Department of Kenyatta National Hospital and analysed.

The patients had been seen at the Neurosurgical Outpatients Clinic having been referred there from other hospitals or from other clinics. Some of the patients had been admitted to the Neurosurgical Unit.

Information was obtained from the files using the outline of Appendix 1. The pattern of data extraction was also applied to the patients studied prospectively.

RESULTS

A total of 27 patients diagnosed as having craniovertebral anomalies over the period 1981 - 1987 were studied. These patients were managed at Kenyatta National Hospital. Most of them had been referred from District or Provincial hospitals.

The number of patients in each year of study is illustrated in table 3.

Table 3: Number of patients presenting in the different years.

Year	No. of patients	%
1981	8	30
1982	1	3
1983	4	15
1984	3	11
1985	5	19
1986	4	15
1987	2	6
TOTAL:	27	100

Patients who were being investigated or awaiting surgery were not included in the study.

A problem with records was that some patients who had been operated upon as shown by the main theatre register but whose files could not be traced. As such, the number of patients with these anomalies is higher than indicated.

SEX DISTRIBUTION:

Of the 27 patients studied 10 were male and 17 female. This is depicted by the histogram below:

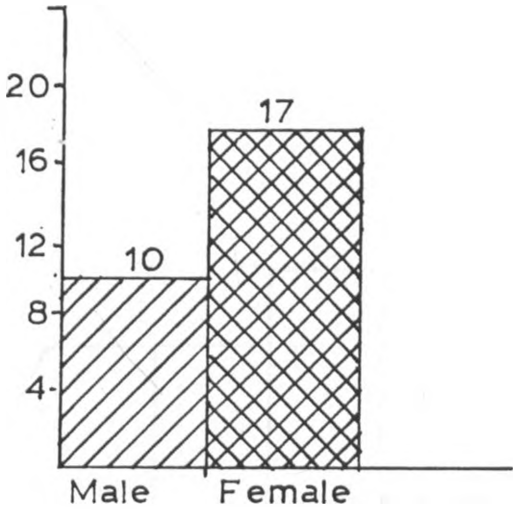

fig 8: Sex distribution of patients.

This gave a male:female ratio 3:5

AGE DISTRIBUTION

The age distribution is shown below in Table 4.

Table 4: Age distribution.

Age range/years	Male	Female	Total	Percentage
0 - 9	0	3	3	11
10 - 19	2	2	4	15
20 - 29	3	2	5	19
30 - 39	2	3	5	19
40 - 49	2	4	6	22
50 - 59	1	2	3	11
> 60	0	1	1	3
TOTAL:	10	17	27	100

This may be more readily visualised by a histogram (fig 9).

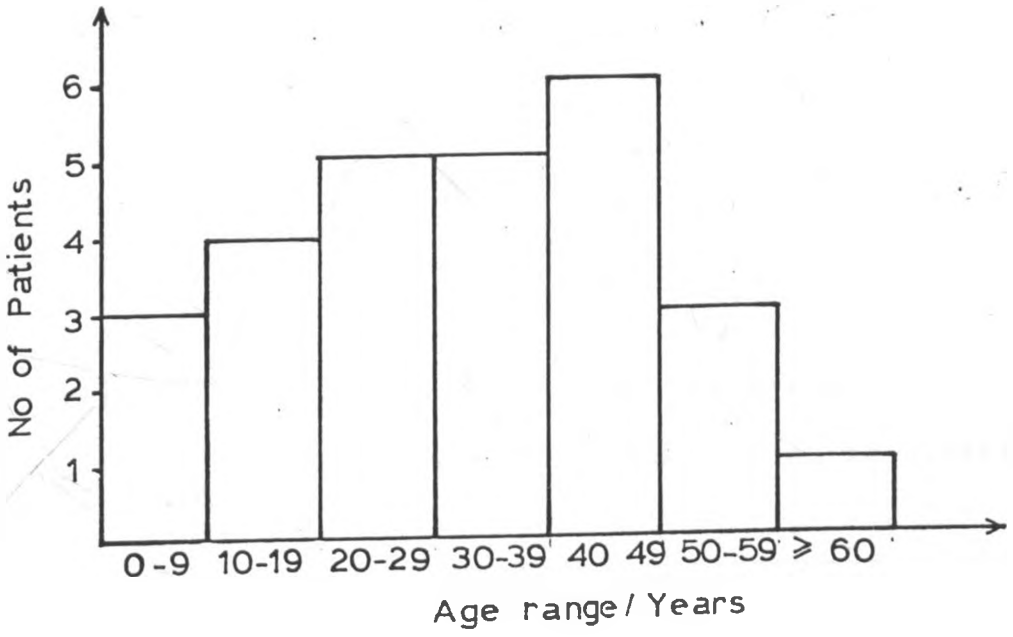

Fig 9: Histogram showing the age distribution.

There was a peak at 20 - 50 years, representing 60% of the patients.

The youngest patient was a 4 1/2 year old girl, while the oldest was more than 60 years old.

The mean age was 31.6 years and the median was 32 years. Females dominated under 20 years of age with no male presenting under 10 years.

TRIBAL AND GEOGRAPHICAL DISTRIBUTION

Ethnic distribution is shown in table 5.

Table 5: Tribal distribution of the patients

Tribe	Number of patients	Percentage
Kikuyu	22	81.5
Meru	2	7.4
Akamba	3	11.1
Others	None	0.0
TOTAL:	27	100.0

All the patients studied were from three tribes, namely, Kikuyu, Meru and Akamba, with the Kikuyus forming the vast majority - 81.5%. These are closely affiliated Bantu tribes from the central part of Kenya.

Their distribution by district is shown in table 6:

Table 6: Distribution of the patients according to district of origin

District	Number of patients	Percentage
Murang'a	13	48
Kirinyaga	4	15
Kiambu	3	11
Nyeri	2	7
Meru	2	7
Machakos	2	7
Kitui	1	4
TOTAL:	27	100

60% of the Kikuyu patients came from Murang'a District. The districts shown in table 6 fall within two provinces, namely, Central and Eastern.

The above distributions are peculiar considering the tribal percentage admissions into Kenyatta National Hospital and the tribal populations as according to the 1979 population census. This is shown by table 7.

Table 7: Kenya tribal distribution and relative tribal admissions into KNH

Tribe	Percentage KNH admission rate (1984)	Percentage of Kenya population (1979)
Kikuyu	33.9	20.1
Akamba	19.8	10.9
Luhya	9.2	13.9
Luo	17.2	13.9
Others	19.9	41.2
TOTAL:	100.0	100.0

CLINICAL FEATURES

SYMPTOMS

The symptoms varied widely with a complex order of progression. They have been summarised in table 8.

Table 8: Symptoms

Symptom	Number of patients	%age
Weakness and ataxia	25	93
Numbness and pain	7	26
Neck pain and stiffness	10	37
Occipital headache	6	22
Visual disturbance	1	4
Nasal voice and regurgitation of food	2	7
Incontinence of urine	2	7
Incontinence of stool	1	4

4 patients (15%) gave a history of trauma and attributed their symptoms to the trauma. In one patient, a grand multiparous woman with 8 children, the symptoms got worse after she gave birth to twins.

Weakness and/or ataxia of the limbs was the most frequent complaint, occurring in 25 (93%) of the 27 patients studied.

Of the 7 patients with numbness or pain in the extremities, both upper and lower limbs were involved.

The variation in the duration of symptoms is shown in table 9.

Table 9: Duration of symptoms.

Duration of symptoms before presentation to hospital	Number of patients	Percentage
0 - 1 (0 - 11 months)	15	55.5
1 - 2 (12 - 23 months)	3	11.5
2 - 3 (24 - 35 months)	5	18.5
3 - 4 (36 - 47 months)	0	0.0
4 - 5 (48 - 59 months)	3	11.5
> 5 (> 60 months)	1	3.5
TOTAL:	27	100.0

15 patients presented within one year of onset of symptoms. This accounted for 55.5% of the patients. Of these, 10, (37% of the total) presented in less than 6 months. The longest duration was 6 years in a 54 year old man from Meru. The mean duration of symptoms was 17 months.

SIGNS

The physical findings were grouped and tabulated, table 10.

Table 10: Physical signs

Physical sign	Number of patients	%
Short neck and low hairline	12	44
Limitation of neck movements	9	33
Long tract deficit	25	93
Ataxia	13	48
Nystagmus	5	19
Bulbar signs	3	11
Posterior column signs	1	3
Muscles wasting	6	22

Long tract deficit comprised upper motor neurone signs or signs of pyramidal system impairment, and included weakness, hypertonia hypereflexia and upgoing plantars. Long tract deficit was the commonest neurological finding, elicited in 93% of the patients.

Bulbar signs occurred in two patients. In one, there was left sided 7th, 9th, 11th and 12th nerve palsies. The other presented with nasal regurgitation of food and had a spastic tongue with fasciculations.

Muscle wasting in the 6 patients was that of the thenar and hypothenar muscles. One, in addition, had wasting of the muscles of the forearm.

CLINICAL DIAGNOSIS

Most patients were referred to Kenyatta National Hospital from various Provincial and District hospitals. The initial diagnoses made by the referring doctors or the doctors first seeing the patients in the various clinics is shown in table 11.

Table 11: Various diagnoses

Initial diagnosis	Number of patients	%age
Brain stem or posterior fossa tumour	6	22.2
Syringomyelia or syringobulbia	1	3.7
Foramen magnum lesion or cervical cord compression	6	22.2
Cervical spondylosis	3	11.1
Cervical spine injury	2	7.2
Meningitis	3	11.1
Craniovertebral anomaly	6	22.2
TOTAL:	27	100.0

A correct initial diagnosis of craniovertebral anomaly was made in 22% of the patients. Other diagnoses such as foramen magnum lesion or cervical cord compression only pointed to the site of pathology rather than its nature.

METHODS OF DEFINITIVE DIAGNOSIS

The diagnostic methods for craniovertebral anomalies are mainly radiological. The investigations carried out were:
plain X-Rays of the skull and cervical spine, tomography of the craniovertebral junction, myelography, angiography and ventriculography.

For the greater part of the period of the study, computerised tomography had not been introduced in Kenya and even after its introduction to Nairobi's private hospitals, none of the patients studied was subjected to it.

The various investigations done aimed at making a definitive diagnosis are summarised in table 12.

Table 12: Various radiological investigations and the number of patients subjected to them.

Diagnostic procedure	Number of patients	%age
Plain X-Rays of head and neck	27	100
Tomography of craniovertebral junction	14	52
Myelography	11	41
Angiography	3	11
Ventriculography	2	7
Encephalography	0	0
Computerised tomography	0	0

All the patients studied had plain X-Rays of the head and neck taken. From these plain X-Rays, a definitive diagnosis of craniovertebral anomaly was made in 15 patients (56%).

In 8 patients (30%) plain X-Rays of the head and neck was the sole radiological diagnostic examination.

As for the other patients where the diagnosis of craniovertebral anomaly was made or suspected on plain radiography, confirmation with tomography or myelography was done.

As for some of the patients with no additional examination other than plain radiography, these investigations had been requested but the myelography and tomography machines were out of order.

Ventriculography was done in 2 patients and was normal. This was in patients with an initial diagnosis of brainstem tumour and posterior fossa tumour.

Carotid angiography was done in 3 patients and was normal. There was really no indication for this investigation. If, as was the case, brain stem or posterior fossa tumours were suspected, then a vertebral angiogram, should have been the relevant initial examination.

Air myelography or encephalography, a useful examination in detecting soft tissue lesions in the cord was not done on any of the patients.

Therefore, the diagnostic examinations of value were: plain radiography, tomography and myelography of craniovertebral junction.

DIAGNOSIS OF THE VARIOUS CRANIOVERTEBRAL ANOMALIES

The diagnoses of the specific craniovertebral anomalies were made on radiological grounds. The number of patients presenting with the various anomalies is shown in table 13.

Table 13: Diagnosis of various craniovertebral anomalies

Radiological diagnosis	Number of patients			
	Male	Female	Total	%
Basilar invagination	4	7	11	41
Occipitalisation of the atlas	8	12	20	74
Chronic atlanto-axial dislocation	2	7	9	33
Others:				
Klippel-Feil Syndrome	2	1	3	11

Most of the patients had combination of craniovertebral anomalies. This was found in 19 (70%) of the patients. 3 patients had basilar invagination with no other bony abnormality.

Only 4 patients had occipitalisation of the atlas alone, while 2 patients had atlanto-axial dislocation alone. Occipitalisation of the atlas was the most common diagnosis, made in 74% of the patients.

3 patients had fusion of two cervical vertebrae. In two this was at $C_2 - C_3$ and in the other $C_4 - C_5$. These three patients had occipitalisation of the atlas, with one in addition having atlanto-axial dislocation.

MODALITIES OF THERAPY

Therapy was either conservative or operative:

Conservative: 12 patients (44%)
Operative : 15 patients (56%)

Of the 12 patients treated conservatively 1 was scheduled for surgery but was lost to follow up and surgery was never done. The remaining 11 patients were prescribed physiotherapy with or without cervical collar. The various reasons for conservative rather than operative therapy are summarised in table 14.

Table 14: Reasons for conservative therapy

Reasons for conservative therapy	Number of patients
Scheduled for surgery but lost to follow-up	1
Non-progression of symptoms	3
Unlikely to benefit from surgery on account of severe disability	7
Unlikely to benefit from surgery on account of old age (more than 60 yrs)	1
TOTAL:	12

Most of the patients treated conservatively were subsequently discharged to their Provincial or District hospitals. Most were lost to follow up.

OPERATIVE TREATMENT

15 patients were subjected to surgery which comprised suboccipital craniectomy and laminectomy of C1 and in most patients C2 as well.

The average age of the operated patients was 31 years. The youngest was 5 years and the oldest 47 years.

In one patient with occipitalisation of the atlas and $C_2 - C_3$ fusion, cervico-occipital fusion was done using cortical bone from the iliac crest and anchored in place by stainless steel wire.

At operation, the following findings were made, not diagnosed pre-operatively:

Arnold Chiari malformation: 6 patients (40% of operated patients)

Bands or adhesions at craniocervical junction: 9 patients (60% of operated patients)

Some patients developed post-operative complications namely hydrocephalus (1 patient), neurenteric cyst (1 patient) and respiratory failure (1 patient).

The patients with hydrocephalus and neurenteric cysts were successfully treated with a ventriculoperitoreal shunt. The patient with respiratory failure was still in the Intensive Care Unit on a ventilator four months after operation.

The outcome of surgery is summarised in table 15.

Table 15:: Outcome of surgery

Outcome of Surgery	Number of Patients	percentage
Improved	10	67
No change	0	0
Deteriorated	4	27
Died	0	0
No follow up	1	7
TOTAL:	15	100

Improvement occurred in 67% of the operated patients. In others the nature of the improvement was not detailed. In some improvement is specified such as a decrease in ataxia, gain of power, cessation of occipital headaches and neck stiffness.

There was deterioration in 4 patients with increasing weakness, ataxia, headache, and neck pain.

No deaths were recorded.

CASE SUMMARIES OF THE PATIENTS STUDIED PROSPECTIVELY

CASE 1:

R. W., a 41 year old female from Murang'a admitted on 9/2/87 having been referred from Murang'a District Hospital with a diagnosis of cerebral tumour with right hemiparasis.

The patient complained of occipital headaches, pain, numbness and weakness of the legs especially the right, and ataxia for a duration of 5 years. The symptoms were progressive and had became worse a few years earlier after delivery of twins. She gave no history of associated trauma.

She had a short neck, low hairline and limitation of neck movements (could not touch the shoulder with the chin). She had normal muscle bulk but marked hypertonia, hypereflexia with ankle clonus. Power was reduced in all limbs. She had no cerebellar signs. She had partial loss of position sense in the legs.

Plain X-Rays of the head and neck and tomography of the craniocervical junction were done from which a diagnosis of

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She had a short neck, low hairline and limitation of neck movements (could not touch the shoulder with the chin). She had normal muscle bulk but marked hypertonia, hypereflexia with ankle clonus. Power was reduced in all limbs. She had no cerebellar signs. She had partial loss of position sense in the legs.

Plain X-Rays of the head and neck and tomography of the craniocervical junction were done from which a diagnosis of

basilar invagination was made.

Surgery was performed on her and comprised suboccipital craniectomy and laminectomy of C_1 and C_2.

Cerebellar tonsils were found herniated through the foramen magnum.

Post operatively she improved. Headaches and numbness ceased. Gait improved thus she could now walk alone with the aid of a stick. She however remained spastic, but the clonus disappeared.

Her photograph and tomographs are shown in Appendix 2.

CASE 2:

K.K., a 31 year old woman was admitted on 15/10/86 having been referred from Machakos District Hospital with a diagnosis of cervical cord compression.

Her complaints were weakness and numbness of the limbs, faecal and urinary incontinence 3 months prior to admission. There was no history of associated trauma.

On examination she had bilateral thenar and hypothenar muscle wasting. No fasciculations. There was hypertonia and hypereflexia of all the limbs with knee and ankle clonus. The plantars were upgoing. Power was diminished in all the limbs especially the left side. Patient was bedridden having progressively become quadriparetic.
There was patchy blunting of sensation to pin prick and touch.

Plain radiography of the craniovertebral junction appeared normal.

Tomography of this region showed marked deformity of the craniovertebral area with occipitalisation of the atlas. C_2 and C3 were fused. The spinal canal was markedly narrowed.

On assessing the patient, it was decided that she would not benefit from surgery but on the insistence of the patient, surgery was done.

This consisted of suboccipital craniectomy, laminectomy of C_1 and fusion of occiput to C_2 and C_3 using cortical bone graft from the iliac crest.

On reversal of anaesthesia, the patient remained apnoeic and was admitted into the Intensive Care Unit, where she has remained for 4 months now on a respirator.

Her photograph and tomograms are shown in Appendix 3.

D I S C U S S I O N

Craniovertebral anomalies are a common cause of spinal cord compression in Kenya. This is borne out by Stewart [34] who reported 17 cases over a 16-month period. However, in 1973 Harries [35] working in Kenya reported that the condition was rare.

Of the patients studied 63% were female giving a male:female ratio of 3:5. This differs from the results of McRae [5], McRae and Barnum [7], McRae [10], Bharucha and Dastur [36], Greenberg [28] and Stewart [34] whose patients all had a male preponderance.

60% of patients presented between the ages of 20 and 50 years giving a peak at the 3rd, 4th and 5th decades of life. McRae and Barnum [7] had his peaks at the 3rd, 4th and 5th decades of life while Bharucha and Dastur [36] had his at the 2nd and 3rd.

The youngest patient was a 4 1/2 year-old girl with occipitalisation of the atlas. List's [1] youngest patient was a 6 year old boy with occipitalisation and Spillane's [6] was 7 years old.

The oldest patient was more than 60 years old. Spillane's [6] patient was 64 years old and McRae's [5] was a 48 year old man.

The mean age of presentation was 32 years with a median of 32 years. McRae [15] had his average age as 31 years. 7 patients were below 20 years. Spillane [6] in his study of 24 cases had 3 below 20 years. This indicates an earlier presentation in our patients.

All the patients were from three tribes, namely, Kikuyu, Meru and Akamba who live in the central part of Kenya. The Kikuyu dominated by a vast majority of 81.5% with 59% of them from Murang'a District. This is highly significant considering that the Kikuyu constitute roughly 40% of all admissions into Kenyatta National Hospital and 20% of the total population. [52]

This regional occurrence was also noted by List [1] who in a review of the literature noted that most cases occurred in the inhabitants of coastal lowlands of northern Europe (Netherlands, northwestern Germany and Sweden). All of List's patients were of Northern European stock.

Globally, there is an unusually high incidence in India [36,37,38] and also in Ceylon (Sri Lanka) and Thailand. De Barrows et al [39] found a frequent occurrence in the North East of Brazil.

CLINICAL FEATURES

The commonest symptom was weakness and ataxia occurring in 93% of patients. This was also observed by of McRae [5,10]. However O'Connel and Turner [23] found occipital headache to be the commonest symptom which could be triggered by head movements, vibrations, driving, coughing, sneezing and straining in the toilet.

2 patients (7%) had urinary incontinence and 1 patient (3.5%) had faecal incontinence. Bharucha and Dastur [36] had 10 of their 25 cases presenting with sphincter disturbance.

4 patients (15%) gave a history of trauma as precipitating their symptoms although the trauma was not severe.

McRae [57] obtained a history of trauma in only 2 (16%) of his 12 cases. McRae and Barmum [7] had symptoms precipitated by trauma in 6 of their 25 cases.

Trauma precipitated symptoms in 15 (32%) of 40 patients of Bharucha & Dastur [36]. In these patients, an interesting feature was the frequency with which porters carrying loads on their heads were affected. No such observation was made in our patients.

Most of the patients (55%) presented to hospital within a year of onset of symptoms. Of these 37% of the total presented in less than 6 months. The average duration of symptoms was 17 months (= 1 1/2 years).

Spillane, Pallis, Jones [6] had 40% of their patients presenting over 1 - 5 years, before presentation to hospital. 60% of their patients presented with symptoms of more than 5 years' duration.

Long tract deficit was the commonest sign occurring in 93% of the patients. This was similarly borne out by List [1], McRae [5,10] and Bharucha and Dastur [36].

Muscle wasting occurred in 6 patients (22%) and comprised of thenar and hypothenar wasting. Bharucha & Dastur [36] had 26% of their patients with muscle wasting. They noted two patterns of wasting; one a symmetrical and selective affection of the shoulder girdle, pectoral and palmar musculature and the other a unilateral and more generalised wasting of an entire upper limb.

Sensory disturbance occurred in one patient (3.5%) and was a posterior column lesion characterised by loss of vibration and position sense. The other sensory modalities were not disturbed.

Spillane, Pallis and Jones [6] had posterior column signs in 30% of their patients with occipitalisation of the atlas.

McRae and Barnum [7] noted posterior column signs infrequently and suggested that the main damage to the cord was occurring anteriorly, presumably by the displaced odontoid process of the axis, rather than posteriorly by a dural band.

A short neck and a low hairline was observed in 44% of our patients. The rest had a normal appearance. 25% of the patients of Spillane, Pallis and Jones had a short neck and low hairline and 25% had a lateral tilt of the head. In their patients as in ours a short neck could not be correlated with any specific craniovertebral anomaly.

After taking the history and examining the patients, a correct diagnosis of craniovertebral anomaly was made in 22% of patients by the referring doctor or the doctor first seeing the patient.

McRae and Barmuum [7] and McRae [10] similarly found a high rate of error in initial diagnosis, a figure of more than 30%. The most prominent error was multiple sclerosis. Others included hereditary ataxia, syringomyelia and syringobulbia, motor neurone disease, cervical cord tumour, posterior fossa tumour, cervical fracture, cervical disc protrusion, cerebellar degeneration and chronic lead poisoning.

Bharucha and Dastur [36], had their most prominent error in diagnosis being hereditary ataxia.

Radiology

Most of the patients had combinations of anomalies with occipitalisation of the atlas leading by 74%, and followed by basilar invagination (41%). This order of frequency is similar to that of McRae [5]. Bharucha and Dastur [36] had occipitalisation of the atlas as the commonest anomaly followed by atlanto-axial dislocation. Spillane, Pallis, Jones [[6] had basilar invagination as the commonest anomaly followed by occipitalisation of the atlas.

Associated bony anomalies - Klippel-Feil syndrome occurred in 3 patients (11%). Spillane, Pallis and Jones [6] had 8% of his patients with Klippel-Feil syndrome. These patients had a short neck, low hairline and extensive fusion of cervical vertebrae.

Associated anomalies of the neuraxis occurred in 6 patients (22%), consisted of Arnold-Chiari malformation and were diagnosed at operation.

Spillane, Pallis and Jones [6] had associated cerebellar ectopia in 33% of his patients and was commonly associated with basilar invagination.

TREATMENT

12 patients (44%) were treated conservatively. This was mainly due to severe disability. Such patients were deemed unlikely to benefit from surgery.

Bharucha and Dastur [36] treated 20 of their 40 patients conservatively by immobilisation in a plater of Paris, felt or plastic collar. Improvement occurred in 6 cases while the rest remained static.

Wadia [31] advocates conservative therapy only as a temporary measure and recommends surgery for all patients except the very old.

3 of our patients were treated conservatively on account of non-progression of symptoms.

Spierings and Breakman [47] suggested that on account of the risks of surgery, patients with local symptoms such as pain in the neck, shoulders, occiput or stiff neck should have no treatment at all; while patients with symptoms localised to the brain stem such as diplopia, nystagmus, dysasthria, dysphagia and vertigo should be treated conservatively unless the spinal canal is very narrow (less than 13mm).

None of their 16 patients who presented without cord signs and who were treated conservatively developed neurological deficits, an argument against prophylactic fusion, a procedure which severely restricts rotational movements of the head.

Operative treatment was performed in 15 (56%) of our patients.

Spierings and Breakman [47] recommend surgical treatment for patients with progressive cord signs to halt their progression. Patients with transient cord signs after relatively trivial injuries which are likely to recur should also be operated on.

Wadia [37] advocated surgery even at a well developed stage of the disease, as a successful operation arrests the deterioration and may even provide some improvement.

A point of note is that the standard operation of removal of the posterior arch of the atlas and posterior margin of the foramen magnum performed for basilar invagination or occipitalisation of the atlas, may result in a disastrous postoperative haematomyelia if performed for atlanto-axial dislocation without previous traction of the neck or attempt at reduction.

Occipitocervical fusion was performed in one patient. Fusion is indicated in atlanto-axial dislocation [28,31,37,38,40], and for the other craniovertebral anomalies once extensive posterior decompression has been performed and major craniocervical destabilisation results. The limitation of motion resulting from fusion of the occiput to the axis is hardly noticed by the average person. [45]

At surgery, Arnold Chiari malformation was found in 6 patients (40% of operated patients). This had not been diagnosed pre-operatively. Spillane, Pallis and Jones [6] had Arnold Chiari Malformation in 8 out of their 24 patients (33%). These had been diagnosed preoperatively and confirmed intraoperatively in 3 of these patients, The other 5 were diagnosed intraoperatively.

O'Connell and Turner [23] found the association in 3 out of their 5 patients with basilar impression.

In Spillane's [6] experience the extent of cerebellar ectopia, bore no relationship to the degree of basilar impression or to the type of the other related skeletal abnormalities.

Syringomyelia was not noted in any of our patients. Klaus [48] showed that of 164 cases of primary basilar impression 22 cases (14%) had fairly severe syringomyelia. Bull [16] found seven cases with this combination in 20 patients with basilar impression.

Of the 15 patients operated upon, 10 (67%) showed improvement. Bharucha and Dastur [36] reported 11 (55%) improvement of their 20 operated patients. 3 patients (15%) did not improve. There were 4 (25%) deaths.

There were no recorded deaths in the patients studied. This may have been due to stringent criteria for surgery. Long term follow up was not available due to patients being discharged to District or Provincial hospitals or simply falling out from follow up.

The age of those who improved after surgery ranged from 5 years to 45 years with a mean of 25 years. This implies a better surgical result at a younger age.

Unfortunately, surgical intervention is accompanied by risk, and a few years ago it could be very dangerous.

In 1953 Garcin and Oeconomos [49] reported that 14 out of 64 operated patients died.

In 1957 Gatai [50] in a summary of the literature found that 33 cases of 90 patients operated on died. In recent years however, the results have improved markedly. Of 75 patients operated on by Driesen et al [51] only 4 died. Follow-up studies showed a marked clinical improvement in one-third of 43 patients. In another third there was no further progression of the condition and half of all patients were able to work.

C O N C L U S I O N

Any patient with symptoms and signs suggestive of upper spinal cord compression, and if such a patient is either Kikuyu, Meru or Akamba, then the presence of craniovertebral anomaly should be suspected even if a history of trauma is present.

Once a definitive diagnosis of craniovertebral anomaly is made and the symptoms and signs of the patient are progressive, surgery should be undertaken with the aim of arresting the clinical course and often reversing the symptoms and signs.

Once the clinical course is allowed to advance, the outlook will be bad whatever form of therapy is instituted.

It is felt however, that despite the marked neurological disability in advanced cases, the offer of surgery may benefit a few. An intensive care unit with facilities for respiratory support should be available when operating on such patients. This appears preferable to condemning these patients to a life of progressive neurological deterioration and death.

PATIENT DATA SHEET

Name

Unit Number

Age

Ward

Sex

Tribe

District of Origin

Date of admission

Date of discharge

Symptoms (onset, duration and order of progression)

Past medical history

Main signs

Investigations;

General inv. Hb & ESR
WBC
Urea
Electrolytes K+
Na+
Ca++
Chest X-Rays

Special Investigations:

Plain X-Rays of skull and cervical spine
Tomography of craniocervical junction
Myelography of cervical spine - positive contrast
- negative contrast
Angiography (vertebral carotid)
Ventriculography
Encephalography (air)
Computerised tomography

<u>Modalities of therapy</u>

Conservative

Operative

Complications of Surgery:

<u>Outcome of Therapy</u>

Improved
No change
Deteriorated
Died

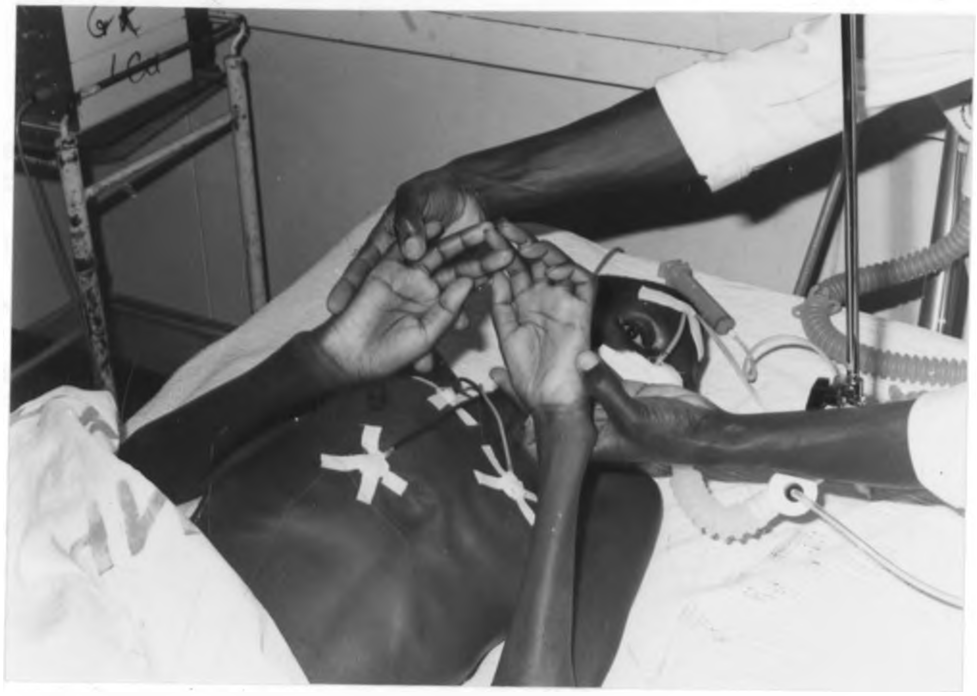
Comments:

APPENDIX II

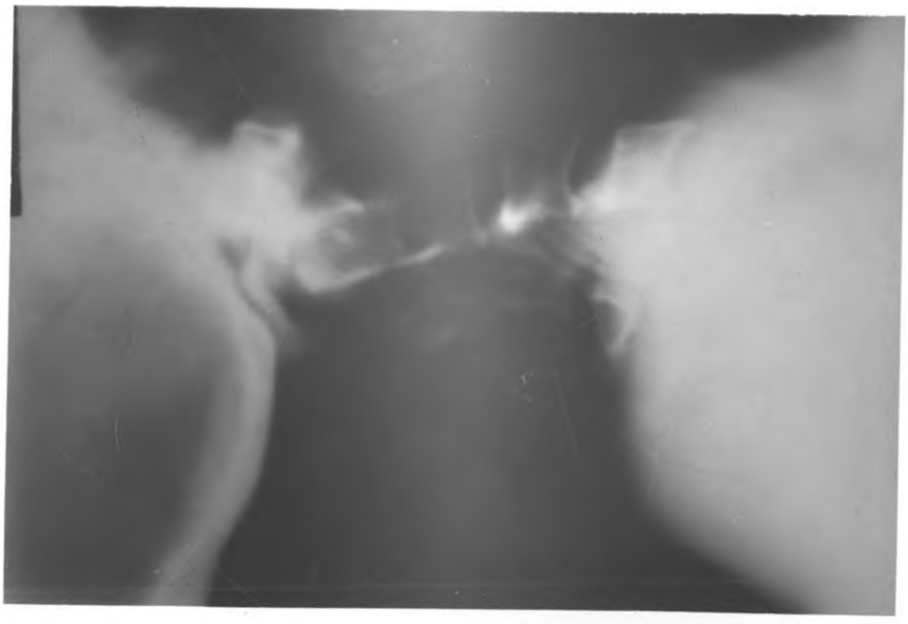
Photograph of RW, a 41 year old woman with a short neck, low hairline and limited neck movements.

Craniocervical junction tomograms of RW showing basilar invagination.

APPENDIX III



Photographs of K.K., a 31 year old woman in the Intensive Care Unit on a respirator. She is quadriparetic. Note the marked bilateral thenar and hypothenar muscle wasting.



Tomograms of K.K.s craniocervical region showing occipitocervical fusion of the antererior arch of the atlas and fusion of the second and third cervical vertebrae.

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