

In the early part of this New Year, I would like to give a second life to this Bulletin in changing somewhat the content and answering some of the comments and criticisms that I have received from some of you. Our last meeting has not been a success insofar as the attendance is concerned and I realize how difficult it is to fill up these few pages with interesting material.

Also, many of you would like more systematization of our presentations. In order to achieve this, I have planned to cover more extensively two subjects, and to give only a short summary of the others. The first subject is an extension of Gus O'Gorman's presentation of two cases of vertebralization of the occiput: with this material as the center point, I will try to complete this aspect of cranio-vertebral anomalies.

The second subject takes advantage of some of our correspondants' experience and is presented in the form of a letter to which we expect more than the correspondant's answer alone. I hope that this first issue of 1981 will please the readers and draw more comments than in the past.

I take this opportunity to express my best wishes for a happy and successful New Year to all good friends of our Neuro Study Club.

Denis Melançon

# BULLETIN OF THE NEURO STUDY CLUB

#### THE CRANIO-VERTEBRAL JUNCTION

Much has been said on this subject but it remains often mis-understood. The main physiopathology rests on the fact that poorly differentiated occipital and upper cervical vertebral segments may lead to stenosis of the foramen magnum and upper cervical spinal canal.

Transitional phenomena exist at that level as they exist lower down along the spinal axis. Cranialization of the junction tends to increase the basal angle or flatten the base which in turn increases the forward bend of the medullary junction ( spinal cord and medulla oblongata ). The reverse transitional phenomenon, conversely, tends to decrease the basal angle or straighten the base as well as free the foramen magnum from any possible intrusion of a long odontoid process. The atlas, in both conditions, is the pivot of one or the other transition movement, but plays very little role by itself in the complications of the anomalies.

To substantiate this subject, we reviewed 47 cranio-vertebral anomalies, described on routine skull and cervical spine examination, and, in most cases, further demonstrated by conventional tomography. (tableau on page 4)

In cranio-vertebral anomalies, symptons and signs will occur when the spinal cord and lower brain stem are impinged upon:

- the lower brain stem may be compressed when
  - . the foramen magnum is small:
    - .. congenital smallness
    - .. hypertrophy of the occipital condyles
    - .. long invaginating odontoid
  - . the angle of the medullary junction is increased:
    - .. platybasia
    - .. short basi occiput
    - .. invagination
- the upper cervical cord may be compressed when
  - the upper spinal canal is narrowed by atlanto-axial subluxation
    - .. due to separate odontoid or, more rarely, absent odontoid
    - .. due to laxity of transverse ligaments
    - .. due to arthritis of the atlanto-axial joint

Continued on page 4 ...

### CORRESPONDANCE:

Letter to Dr. Giuseppe Scotti, concerning a case of sequestration of the fourth ventricle.

Dear Giuseppe,

You have studied and written up recently many cases of the so-called "sequestered fourth ventricle". I would like, in the following lines, to present to you a clinical problem of similar nature that we recently studied.

Our patient is a male of 27, who was operated upon in 1975 for a hemangioblastoma of the cerebellum. A tumour and cyst were removed. Following surgery, he developed hydrocephalus which was corrected with a ventriculo-atrial shunt. In the following two years, he frequently developed episodes of double vision, headaches, nausea, vomiting and ataxia. Although blocking of the shunt was suspected, it always seemed to be fonctionning well and computed tomography was not showing hydrocephalus. In May 1978, ventriculography was performed and it indicated that there seemed to be an aqueduct stenosis. In June 1978, following arteriography, his suboccipital craniectomy was reopened and a small nodule of hemangioblastoma removed from the cerebellum. In the following months his shunt was revised many times with some improvement each time, but with recurrence periodically of the same episode of deterioration.

Only at the time of his recent admission did we notice that the fourth ventricle was larger than normal, somewhat ballooned, suggestive of some local inner pressure. It was also noted then that the size of the fourth ventricle had varied somewhat over a period of many months. It was then decided to study this anomaly with Metrizamide Body Scanning following lumbar injection of the contrast and with sequential examination. The following observations were made:

- 1) The large fourth ventricle filled easily with contrast;
- 2) No filling of the posterior third ventricle was seen;
- Persistent filling of the fourth ventricle and basal cisterns was noted (6 hours);
- 4) Poor filling of the convexity sulci was present on the delayed scan as well.

From these observations, we concluded that the fourth ventricle was sequestered, because of a stenotic aqueduct, but not because of incompetence of its foramina; instead, it appeared that its sequestration was shared by the spinal SAS and basal cisterns, due to incompetence of the convexity sulci to carry normally the cerebro-spinal fluid and contrast up to the region of the pacchionian granulations.

We have not read about that observation before and we will appreciate your comments in this matter. I include all films that may be pertinent.





1)

4)



3)



## UNILATERAL OCCIPITAL CONDYLE HYPERPLASIA OR VERTEBRALIZATION

by Gus O'Gorman



Six year old boy presents with head tilt and headache. Conventional tomograms and CT in the coronal plane demonstrate hypertrophy of the left occipital condyle which is thought to represent vertebralization of the occiput on one side.

Incomplete or partial vertebralization of the occipital squama is rare and of no pathological significance. Minor forms of vertebralization of the occipital condyles are relatively common. If vertebralization is more marked the condyle is connected to the skull base only by a bony bridge and on frontal view resembles a peninsula. Wackenheim believes vertebralization of the occiput and especially of the condyles to be the opposite malformation of pathogenic mechanism i.e. narrowing of the spinal canal rather than malformation.

Reference: A. Wackenheim Roentgen Diagnosis of the Craniovertebral region. Springer-Verlag, 1974.

PARACONDYLIC PROCESS

by Gus O'Gorman

This was an incidental finding on a 14 year old boy.

The paracondylic process is a broad based bony prominence arising at the site of the occipital condyle from the exo-occipital bone laterally (part of the occipital bone) and pointing in the direction of the lateral end of the transverse process of the atlas. This particular example shows a false joint with the transverse process of the atlas.

The epitransverse process is found in the same location but arises from the transverse process of the atlas. Both may occur either unilaterally, bilaterally, separately or together on different or the same sides. Genetically, both processes are considered to be part of the transverse process of the pro-atlas.

Reference: Detlef Von Troklus and Walter Gehle The Upper Cervical Spine Grune & Stratton, New York.



## THE CRANIO-VERTEBRAL JUNCTION

But all anomalies encountered can exist without clinical manifestation. The most benign and common anomaly is fusion of the atlas to the occiput.

The manifestation of vertebralization (occipital vertebra) usually has no clinical counterpart. As a matter of fact, it tends to straighten the clivus and thus straighten the medullary junction (medulla oblongata and spinal cord). Also to separate a possible long odontoid from the foramen magnum (stenosis) or the basi-occiput (impression-invagination and platybasia).

Of the 47 observations, 18 had some clinical correlation but only 13 were retained as significant: 6 neck pain - 3 myelopathy - 3 lower cranial nerves palsies - 1 gait disturbance

- Of the 6 neck pain presentation, all six had an atlanto-axial subluxation.
  - .. 3 due to a separate odontoid
  - .. 2 due to joint disease (rheumatoid arthritis)
  - .. 1 due to laxity of ligaments (association with occipitalization)
- Of the 3 myelopathies,
  - .. I was due to stenosis of the foramen magnum (hypertrophy of condyles)
  - .. 2 were due to atlanto-axial subluxation (1 because of a separate odontoid)
  - Of the 3 lower cranial nerves palsies,
  - .. all 3 were due to marked basilar impression
  - .. 2 due to a short basi-occiput
  - .. I due to a long odontoid (with AA subluxation and rheumatoid arthritis)
- . The presentation of gait disturbance was due to a complex anomaly, but mainly to an atlanto-axial subluxation secondary to an absent odontoid.

Of the 29 incidental presentations - 21 were anomalies of the atlas . 10 fusion . 6 hypoplasia

### MISCELLANEOUS

Among the other cases presented at the December Study Club were:

- . A case of Spinal Arterio-Venous Malformation discussed by Garry Bélanger.
- . A case of chronic subdural fluid collection likely following myelography, in a patient who had been complaining of postural headaches since his myelography several months before presented by Denis Melançon.

### THE LOGO

You have probably noted a change in the logo of Neuro Image from number one to number two issue.

I have chosen the present one, "a slice of the brain misformatted by the computer", for its many analogies:

A rising star at the horizon, to which leads a curved road: A rising new technology for the disclosing of the brain structures, and its uneasy approach.

An indirect approach to an imperfect slice of the brain: The relative imperfection of present technologies and the careful approach they require.