

GENERAL STAFF MEETING  
UNIVERSITY HOSPITALS  
UNIVERSITY OF MINNESOTA

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## I. PEOPLE

1. Oncologist William Thomas "Bill" Peyton pondered over growing indifference to Tumor Conference held every other week at 11:00 o'clock on Friday, called a meeting last week of representatives from each service to consider the problems of the Tumor Service. One wrote, some came, others sent regrets, all present discussed the subject. Outstanding impressions -- Opened in November 1925 and called the Cancer Institute, it is gradually and persistently living up to its name. Signs of the times - rapid turn over of hospital patients (next to obstetrics and newborn), rapidly growing Out-patient service, excellent follow-up, systematic treatment of the various forms of the disease, almost complete utilization of special treatment facilities, growing list of publications.

Apparent to all is lack of special interest in the disease on the part of many, natural interest by a few. Growing pains of the loosely organized group indicate that an organization "consultation" is becoming more necessary as time goes on. Some day the "Memorial Plan" will be put into effect, placing the responsibility on one man's shoulder for the treatment of one special form of the disease in both the Hospital and Out-patient Department. Through necessity, this will not be a short term arrangement, or limited to the full-time group. Under these key men, fellows, internes and clerks will get better services. Until then, each service will through necessity cooperate with the Out-patient Tumor Clinic by giving each malignancy patient on leaving the institution a special appointment slip for a return visit. Services, so far as they desire, will be represented in this clinic, others who have no special representative will refer their patients to the group for follow-up.

Unique is the problem of cancer in that it crosses clinical-anatomical boundary lines and segregates for study all cases on a pathological basis. Private hospitals are asked to appoint cancer committees to supervise the treatment of all cases of malignancy in their institutions. Surely, a Cancer Institute should do as much. Short course graduate students possibility of exhausting special treatment facilities and the need for a hospital procedure book (also discussed). Dean Scammon said in part "that he had heard many of the problems discussed today de-

cidated at previous meetings. Until such time as all of our work is organized "on paper", our problem will be decided over and over again". He predicted that within a week few would be able to remember what had been said and some would even misquote the statements made. Result: He was misquoted within ten minutes, another meeting in which departmental heads will be present is to be called, some day the service will be organized on the Memorial basis. Remarkable is the progress which has been made. All credit is due the men who have brought this about. It has been due to their tenacity and thoroughness that the Cancer Service has become the problem that it is. No other university offers its students so close an arrangement with an active Cancer Service, Thanks to the acuity, vision and generosity of the Citizen's Aid Society.

2. Radio-fan extraordinary Paul Richard Snyder, Preston, Minnesota, wrote as usual Wednesday afternoon, died Thursday afternoon. For the past three years, ex-Marine, Horticulturist, College Graduate Snyder has faithfully listened each week to the broadcast of the Minn. State Medical Association from WCCO on Wednesday at 11:15 A.M. Every split infinitive was noted, poor delivery pointed out, good subjects applauded and poor ones booed. Between times he managed to paint a word picture of the disease from which he suffered - disseminated sclerosis. Although he never failed to enjoy the humorous part of our existence, there were many times when he was not "euphoric". Interesting games occupied his time. Thoroughly conversant with the ultimate outlook of his trouble, he at one time arranged in the order of frequency the possible manner of his death, completed his funeral arrangements and formed a club of patients suffering with his disease. Disseminated sclerosis claimed in the tenth year of its existence, the chief critic and guide of the Minnesota State Medical Association's Broadcasting Service.

3. Charles Peyton Rufe arrived in Chalfonte, Pennsylvania February 29, 1932. This triple threat young man starts life under auspicious circumstances Born of distinguished parents, Dr. and

Mrs. H. Redding "Fritz" Rufe, named after one of the future immortals, born on Leap Year Day much will be expected of him. Congratulations from all to our good friends - the Rufes.

The Superintendent requests that the rule prohibiting smoking in the corridors be strictly observed. Also, that as a simple matter of courtesy that consultations be requested in the name of the chief of the service or one of his associates. Thank you.

## II. ABSTRACT.

### THE SCIATIC SYNDROME

Grossman and Keschner.

Arch. of Neur. and Psych. 21:398-411, 1929.

Ever since Cotugne described sciatica, this term has been loosely applied to any condition characterized by pain in the lower part of the back and lower limbs, regardless of the underlying cause and of the other clinical manifestations associated with the pain. One finds the term "sciatica" applied to designate such conditions as sciatic neuralgia, sciatic neuritis, sciatic radiculitis, meralgia paresthetic, lumbago, myalgia, myositis, and what not, as if all these conditions were one and the same clinical entity. As pain in the lower back and in one or both limbs, is in the majority of cases merely a symptom-complex which may be present in various and different clinical entities, we would suggest that this symptom-complex be designated as the "sciatic syndrome."

It was for these reasons that the authors undertook this study. They had at their disposal 317 cases of this syndrome admitted to the neurologic wards of the Mount Sinai Hospital during the past 10 years.

An analysis of the material permitted the following grouping:

1 Patients with paroxysmal lancinating pains in the lower part of the back and in the lumbosacral distribution without organic neurologic signs and without evidences of involvement of the osseous-arthritic structures or of their adjacent soft parts, conditions which they designate

"sciatic neuralgias," 3 cases.

2 Patients with more or less continuous pain in the same distribution with evidences of involvement of the osseous-arthritic structures or their adjacent soft parts. This group includes 267 cases and is subdivided into two groups:

a Patients without neurologic signs of involvement of the lumbosacral roots or nerves. For the sake of convenience they designate these "orthopedic cases."

b Patients with objective signs pointing to involvement of the lumbosacral roots or nerves -- radiculitis or neuritis.

3 Patients in whom the sciatic syndrome is an early and prominent feature of a clinical picture the underlying cause of which is a neoplastic, inflammatory, vascular or degenerative disease of the spine, pelvis, cord, meninges, roots or nerves in this area- 47 cases.

As a result of this study the authors came to the following conclusions:

1. The term "sciatic," as it is generally employed, is misleading. It tends to direct attention away from the most common and remediable underlying cause of the various clinical conditions characterized by pains in the lower part of the back and limbs. The use of this term should therefore be discontinued, and an effort be made to determine the underlying cause and to designate the condition accordingly, and as being associated with the sciatic syndrome.

2. In the absence of a polyneuritis or polyradiculitis from whatever cause (toxic, infectious, metabolic, constitutional, vascular, etc.), a primary mononeuritis or radiculitis of the sciatic nerve is extremely rare. In practically all of our cases the sciatic syndrome was secondary to some pathological process in the osseous-arthritic structures or their contiguous soft parts in the region under discussion.

3. Cases in which the patients have paroxysmal attacks of lancinating or shooting pains in the lower part of the back and in the lumbosacral distribution without objective evidences of

involvement of the lumbosacral roots or nerves, or of the osseo-arthritis structures or their adjacent soft parts (muscles, tendons, fascia, etc.) in this region should be designated as "sciatic neuralgia." The presence of only 3 cases in a series of 317 indicates the rarity of this condition, notwithstanding that most authors emphasize its frequency and employ the term "sciatica" as if it were synonymous with "sciatic neuralgia."

4. By far the largest number of patients presenting the sciatic syndrome show demonstrable evidences of involvement of the osseo-arthritis system or of the contiguous soft parts in this region. This group may be subdivided into two subgroups: (a) Cases without demonstrable signs of organic nervous disease. In the absence of such signs, these cases present an orthopedic problem and would, for the sake of convenience, be designated as "orthopedic cases." We would also emphasize the importance of early and proper treatment of these patients lest the pathologic process in the osseo-arthritis system and the adjacent soft parts become more advanced and involve the nerve roots or nerves themselves in this region. (b) Cases with demonstrable evidences of involvement of the lumbosacral roots or nerves. Depending on the nature and distribution of the involvement of the nerve we would designate these as cases of lumbar, lumbosacral or sacral radiculitis or neuritis. This subgroup includes 64 per cent of our entire material.

5. There are a certain number of cases in which the sciatic syndrome may appear as an early and prominent feature of a clinical picture the underlying cause of which is a neoplasm of the spine, pelvis, cord, meninges or roots. The fallacy of designating these cases as "sciatica" is self-evident.

6. Except in such rare cases as sacralization of the fifth lumbar vertebra, calcification or ossification of the soft parts, or in primary or metastatic, neoplastic, or tuberculous processes in the spine or pelvis, positive x-ray observations of spondylitis or arthritis are not conclusive as to the etiologic relationship of these observations to the sciatic syndrome.

EXTRACT: From Teaching Outlines in Neurology and Psychiatry (J.C. McK). Illustrative cases of type 3 follow (neoplastic sub group).

### III. CASE REPORT

#### SCIATIC SYNDROME: SACRAL CHORDOMA . Pearson.

The case is that of a white female, 50 years of age, admitted to the University Hospitals 1-28-32 and discharged 2-13-32 (16 days).

#### 18 Months Ago (Back Pain)

8 - -30 - Patient noted lameness in back. Pain was of chronic nature and occurred during the day and night.

From this time until the middle of November 1931, patient's condition was about the same. She was treated for neuritis during this time by various physicians and a chiropractor. Some relief was obtained by the chiropractic treatments.

#### Coccyx Removed, left pain

10-13-31 - Patient was operated upon, and the coccyx removed. Directly following this, the patient began to have pains in her left leg of the nature of a dull pain below the knee.

#### Bilateral Pain, Sphincters

1-1-32 - Patient began to have pain in right leg. She was given two injections into the sacrum for relief of sciatic pain. These pains have returned again. She has had some difficulty in moving bowels since August 1930, and this has been getting progressively worse. Patient had difficulty in passing urine, and on a few occasions it was necessary for a catheter to be inserted.

#### Tumor

1-28-32 - Admitted to the University Hospitals. Physical Examination shows a white female in recumbent position and in very marked pain. Patient is somewhat emaciated. Abdomen - liver and spleen are not palpable; tenderness is present in both lower quadrants. There is a mass palpable in the sacral region which is tender to pressure; otherwise the back is normal. Laboratory: Urine - specific gravity 1.021. Blood - Hb. 86%, wbc's 9,550, Rbc's 700.



L 24%, M 4%. Wassermann - negative. Progress: Pulse and temperature normal.

#### X-RAY

1-29-32 - X-ray - sacrum and coccyx, chest - There is a calcified Ghon's tubercle in the right middle lobe. No other evidence of disease in the chest. There is complete absence of the coccyx as the result of a previous operation. In addition, there is almost complete wiping out of the entire structure of the sacrum, this being particularly marked in the upper and posterior portions. The sacro-iliac joints are fairly intact as is the lumbosacral joint. There is a small fairly densely calcified area in the middle of the area of the destruction which may represent some bone debris or possibly some calcification secondary to the destructive process. There is no other evidence of new bone formation. The entire appearance is most suggestive of a metastatic malignancy, for example carcinoma of the breast or nephroma. The possibility of a primary tumor must also be borne in mind. Conclusions: Calcified Ghon's tubercle, right. Operative removal of coccyx. Malignancy in sacrum, probably metastatic. Morphine sulphate gr. 1/4 given. Complains of pain in back. Neurological examination done and the following additional history elicited given. About 1 1/2 years ago, patient experienced severe pain in the lumbar region. One year ago, patient noticed numbness about the rectum, so it was necessary for her to strain in order to pass urine. She would occasionally have an involuntary bowel movement. Catheterization was necessary on two occasions. About five months ago, she became weaker and her condition seemed to be worse. About three months ago, the coccyx was removed. About three weeks ago, she had severe pain down the right leg. Patient has not walked since October 1931. Examination - cranial nerves - right pupil is smaller than left. There is a limitation of motion of the lower extremities due to pain. Patient is in recumbent position and cannot walk. There is a perineal area of anesthesia typical of caudal portion of cord involvement.

#### Deep Therapy

2-1-32 - Pyramidon gr. x. Ammonium chloride gr. xx. Urtropine gr. x. given three times daily. Catheterized, 200 c.c. urine obtained. Urine - contains some

sediment, is very dark. Complains of severe pain in back. Pyramidon gr. x. Morphine sulphate gr. 1/4. 110% skin erythema dose to anterior lower abdominal portion and 100% skin erythema dose to skin over posterior, lumbar and sacral regions, each in two treatments during 2-3 to 2-13-32. This was suggested and given.

#### Biopsy

2-10-32 - Medical note: Patient has had two x-ray treatments and feels fairly comfortable. Second treatment caused much less of a reaction than the first. A biopsy was taken from which a diagnosis was made of chordoma. Clinical diagnosis was also chordoma.

#### Better

2-13-32 - Patient had a deep x-ray treatment. M.S. gr. 1/6. Patient feels very well.

#### Discharged.

To be followed through Out-patient Tumor Clinic (mail or in person).

#### Note:

Examination of sections of small piece of tissue from sacral tumor shows a most interesting picture. There is a dense, fibrous tissue framework throughout. The meshes are filled with round and spindle shaped cells which have deeply stained nuclei and show many mitoses. In the background so to speak, there are nests of vacuolated round, polyhedral cells with clear cytoplasm. The general appearance is that of a rapidly growing cellular tumor which should be very radiosensitive. The histological clue of its possible origin is to be found in the small nests of cells characteristically seen in chordomas. In our experience rapidly growing chordomas may assume the structure of a sarcoma.

#### IV. CASE REPORT

##### SCIATIC SYNDROME, SACRAL CHORDOMA.

Path. Pearson.

The case is that of a white male, 53 years of age, admitted to the University Hospitals on 8-18-31 and died 10-4-31 (47 days).

#### Sciatic Syndrome

5-25-31 - Patient noticed soreness over his lower back. He had this constantly

for 3 or 4 days. Later on, the pain went down the posterior aspect of the right thigh almost to the knee. He described the pain as being a recurring type of gnawing pain which would last about an hour, usually relieved by application of heat. He went to a physician who diagnosed the case as sciatic rheumatism. He gave the patient some codeine pills which relieved the pain somewhat. Continued this way for three months.

#### Walking Difficulty

8-1-31 - Patient stated that he had difficulty in walking because of a drawing of the tendons of the back of the knees together with pain in his calf muscles. These severe pains lasted about 4 or 5 days and would come on at intervals of 3 or 4 hours. He was unable to lie on his right side because of the soreness. The right hip was somewhat swollen and very tender to pressure.

#### Sphincters

8-10-31 - Patient noted numbness in the right foot which lasted about a week. He also had difficulty in starting the urinary stream. During the last three months he has lost about 27 lbs. in weight.

#### Hospital (after 3 Mo.)

8-18-31 - Admitted to University Hospitals. Physical Examination reveals a well-developed and poorly nourished, white male. The mucous membranes are somewhat pale. No deformities are present. Ears - hearing is markedly impaired on the left side. Abdomen - no masses or tenderness present. Extremities - slight amount of atrophy of muscles of both legs; some flaccidity of the right foot; no voluntary motion in right foot at all.

#### Spine

Slight scoliosis to right of upper dorsal spine; no tenderness or masses over entire spine. Neurological examination - some weakness of right leg, especially on abduction; right ankle jerk is greatly diminished; loss of superficial touch and pain on right leg; loss of deep muscle pain and tendon pain on right leg; position sense is normal; Babinski's and Romberg's are negative.

#### Rectal examination

shows a mass near the right border of the sacrum outside of the bowel.

Laboratory: Urine - specific gravity 1.032, occasional rbc's. Blood - Hb. 73%, rbc's not counted; wbc's 3,250, Pmn's 60%, L 37%, M 3%; another specimen - rbc's 3,300,000.

#### X-RAY

Pelvis and lower lumbar spine - There is a fairly sharpened circumscribed and rounded area of destruction in the region of the right sacroiliac joint and involving both the sacrum and the ilium. The bone is almost completely wiped out and there is no evidence of new bone formation. The appearance is fairly characteristic of an osteoclastic tumor such as a hypernephroma. There are several very small rarefied areas in the crest of the left ilium which may also represent metastases although this is rather doubtful. Conclusions: Osteoclastic tumor of right sacroiliac region, probably hypernephroma. Progress: Given pyramidon gr. v, three times daily. Hot water bottle given to right side.

#### Urologic Study

8-21-31 - X-ray - K.U.B. - Neither kidney can be well visualized because of the large amount of gas and fecal matter in the colon. The right kidney appeared to be rather small. The left one could not be made out. Both psoas shadows can be definitely seen and appear to be within normal limits. It is entirely possible that the failure to visualize the left kidney may be due to some pathology present in this region. Suggest re-examination.

#### Laboratory

Urine - specific gravity 1.025, otherwise negative. Spinal fluid - clear, colorless, pressure 150 to 250 mm., no cells, Nonne +, Noguchi heavy +, Colloidal gold 0000133100, Wassermann negative. Stool - benzidine faintly +. N.P.N. - 24.8 mgs. P.S.P. - total 65%.

#### X-RAY

8-22-31 - X-ray - chest - The trachea, heart, mediastinum and both diaphragms are normal. The lungs show no evidence of pathology. Conclusion - negative chest. Patient complains of pain in the right hip.

8-24-31 - X-ray - colon - shows a mass in the right side of the pelvis displacing the rectum to the left. The colon

did not appear to be involved but filled out well throughout its length. There were several areas suspicious of diverticulae. Conclusion: Displacement of colon to left secondary to mass in right side of the pelvis.

Proctoscopic examination - 24 cm. There is a small anal fissure in the posterior midline. Hypertrophied anal papillae, grade I. There is a hard, fixed mass 12 cm. above the anus in the right posterior and right side of the rectosigmoid. Mucosa is intact. This mass is entirely extrarectal. No biopsy specimen could be obtained. Urine - specific gravity 1.005, otherwise negative. Patient has slight backache and pain in the right hip.

#### Repeat Urologic Study

8-25-31 - Skiodan - Plates of the kidneys following skiodan injection were unsatisfactory on account of the barium in the colon. Conclusion: Unsatisfactory skiodan. Patient is quite uncomfortable and has a great deal of pain.

9-1-31 - Pyelogram - The left pyelogram shows a normal ureter and kidney pelvis. There is some evidence of pressure on the right side of the bladder. The area of bone destruction, previously visualized in the sacrum, is still present and somewhat larger than previously. Conclusions: Normal pyelogram, left. Pressure on bladder, right.

Cystoscopic examination - The bladder itself is apparently normal, but of rather large capacity. There was a suggestion of encroachment upon the bladder in the region of the right posterolateral portion of it, coming down slightly in the region of the right side of the trigone. Both ureteral meati were normal, and there were spurts from both sides, (apparently clear). The prostatic urethra was normal except that the right lateral lobe of the prostate was a little large and somewhat encroached upon the prostatic urethra. The left side was catheterized without any difficulty, but although we tried for a long time to pass catheters or filiforms into the right meatus we could not do it. The obstruction was right at the meatus itself. A specimen was obtained from the left side and later a left pyelogram was done. The specimen from the left side did not show any pus.

There were some red blood cells which were probably traumatic. The pyelogram itself on the left was perfectly normal. The cystogram obtained from the reflux of solution injected into the left side suggested a slight filling defect in the right side of the bladder. It was felt that the mass in the right side of the pelvis was entirely extraurinary. However with absence of inflammation on the right side of the urinary tract, there is a remote possibility of a malignancy in the prostate itself, although from all evidence obtainable there is absolutely no indication of the presence of any such malignancy. The incomplete diagnosis is normal left urinary tract, normal bladder and prostate with the exception of slight enlargement of the right lobe of the prostate intraurethrally is made.

#### Skiodan

9-5-31 - Skiodan - The right kidney pelvis is fairly well visualized as is the ureter and shows no particular evidence of abnormality. The right kidney itself could also be seen and does not appear particularly enlarged. The bladder is greatly dilated and well visualized. The left kidney pelvis and ureter could not be visualized. The cause for this could not be determined. Stool - + benzidine. N.P.N. - 20.8. M.G.M. Blood - Hb. 65%, rbc's 3,800,000 wbc's 4,250.

#### Gastro-intestinal study

9-9-31 - The stomach shows marked hypertonicity and there is marked spasm of the pylorus. Some deformity of the duodenal bulb is present and the appearance suggests an old duodenal ulcer. There is no evidence of malignancy in the stomach. Codeine gr. 1 1/2, elixir iron strychnine and quinine drams i 3 times daily. Pyramidon gr. v, three times daily. Complains of severe pain in the back and general discomfort.

#### Rectal

9-12-31 - Genito-urinary consultation Examination shows a hard, nodular mass almost filling the pelvis extending into the right iliac fossa. The mass is fixed to the abdominal wall. Diagnosis is a mass probably metastatic carcinoma of unknown origin or osteogenic sarcoma of the innominate bone must be considered. No biopsy.

Deep Therapy

9-13-31 - Request for deep therapy - 163% given to tumor, right lower quadrant to the pelvis, from four sides in eight treatments (9-14 to 9-30-31).

Neurological examination

9-19-31 - Emaciation, looks older than the age given. Peripheral arteries are hardened. Cranial nerves and nerves of the upper extremity and trunk are negative. There is a marked weakness in the muscle group of the right lower extremity, external rotation of thigh, flexion of thigh, adduction of thigh, extension of the leg and dorsal flexion of foot. There is moderate weakness in the groups named. Decreased knee and absent ankle jerk on right. Very active knee and ankle jerks, left. Negative Babinski's. There is marked loss of touch and pin sensibility on right buttock down the back, right thigh and on the right heel and lateral plantar aspect of the right foot. There is decreased muscle pain and sense in right calf. Absence of vibratory sense in both lower extremities. Conclusion: Paralysis due to pressure on the right sacral plexus.

10-3-31 - Patient seems very drowsy and responds poorly. Pulse 120. Temperature to 100.

Exitus

10-4-31 - Patient is very restless. Complaints of pain. Not very codeine sulphate gr. i given with some relief. Respiration are now 36, labored and shallow. Cheyne-Stokes respirations noted. Patient is cyanotic. Caffeine sodium benzoate gr. 7 1/2 given. Adrenalin i cc. Oxygen tent instituted. Intravenous of 5% glucose and 1000 c.c. saline given. Pulse is very fast and irregular. Respirations are shallow. Skin is cold and clammy. Heat is applied. 8:40 A.M. - Patient expired.

AUTOPSY:

A partial examination was done February 26, 1932 in the Department of Anatomy. The body was sent there because it was unclaimed. The partial examination was conducted through the courtesy of the Department of Anatomy.

A partial dissection of the nervous system in the upper extremity and opening the body cavity had been done. The tumor area is well preserved. The lungs show

anthracosis, congestion and wedge-shaped hemorrhagic areas at the periphery which on section proved to be infarcts. The heart and aorta are without note.

The liver, spleen, stomach and intestines do not show any tumor. The kidneys and adrenals are normal. The bladder wall is slightly thickened. A view of the pelvis reveals an interesting change. There is no visible evidence of tumor tissue. The left half is normal in contour. In the right posterior angle, a marked fullness is seen which results in flattening at this point. The peritoneum is intact over this region. A small nodule is found on the outer surface of the rectum. On section, this proves to be fat.

The right posterior portion of the pelvis is incised, and infiltrating tumor tissue is found. The finger can be passed backward into the defect in the bone. The body is then turned over and the tumor exposed from the dorsal surface. It corresponds with the wiped out area in the sacrum (x-ray) and has a fairly definite border. When opened, it contains dark red, hemorrhagic tissue which is very friable. The medullary cavity is apparently invaded from the external border of the defect.

Sections made of the tumor show that the stroma is fairly abundant. There are many clear areas in the intracellular substance which suggest deposits of mucin. Many thin-walled blood vessels are seen. Several characteristic nests of vacuolated chordoma cells are made out (physaliphorus cells). Many exhibit nuclear hyperchromatism, some are multinucleated, syncytial masses. Hemorrhagic areas and plasma cells are present. Diagnosis: Chordoma of sacrum and ilium (intra and pre-sacral) with invasion of the retroperitoneal pelvis tissues.

V. ABSTRACTS: CHORDOMA

Abstr. Pearson

1. Historical.

(1846) Virchow noted on the surface of the clivus Bumenbachii a small, slimy excrescence attached to a defect in the bone.

(1856) Luschka also described these soft, lobulated, jelly-like masses protruding into the skull from the clivus and perforating the dura.

(1858) Müller described them as



chordal rests because of their characteristic clear cells. He studied a series of older human fetuses and newborn infants and found this tissue in small openings of the clivus, tooth of epistrophius sacrum and developed the anatomic basis for the study of chordoma. In sphenoccipital synchondrosis it remains as a small soft mass analogous to nuclei pulposi of the intervertebral discs which are generally recognized as remnants of chordal tissue. Müller also showed that in the region of the sphenoccipital synchondrosis the noto-chord has a decided tendency to approach the superior surface of the basilar cartilage.

(1894) Ribbert confirmed observations of Müller and was first to suggest name "chordoma". Ribbert placed pathology of chordoma upon sound basis. Virchow, who first noted these masses in clivus, thought they were cartilaginous and gave them the name of ecchondrosis physaliphora. Ribbert also experimentally produced benign chordomas by puncturing anteriorly the intervertebral discs of rabbits.

(1914) Alezais and Peyron demonstrated in great detail histogenesis and evolution of tumor.

(1864) Klebs described the clinical symptoms of clivus tumors and was the first to do so before tumor was really known as chordoma.

## 2. Frequency of the tumor.

Tumors of the noto-chord are probably not as uncommon as reports suggest. Up to 1927, 57 cases were reported, 27 in clivus Blumenbachii, 28 in sacrococcygeal region, 1 in cervical region and 1 in lumbar region. Earlier investigators found ecchondrosis physaliphora of the clivus Blumenbachii was a fairly frequent malformation being found in 3% of the autopsies by Ribbert and 2% by Stewart and Morin. Coenen (1925) in a splendid review collected not only most of the published cases of chordoma up to that time, but also most of the cases of noto-chordal heterotopia (ecchordosis) which had been reported. The latter are without clinical significance being mere casual findings in postmortem room but are of importance as the possible starting point of chordoma. Others: Stewart and Morin in a consecutive series of 350 autopsies especially investigated from this point of view found 1 case in a 36 year old female. As the brain was removed, a soft, flat, transparent, jelly-like structure

disengaged itself from the pons and adhered to dorsum sellae. It was attached by a very slender pedicle to middle of dorsum sellae about one-half inch behind posterior margin of pituitary fossa. At this point there was a small aperture in the dura mater. Also studied 24 published cases of ecchordosis in which age was stated in 15 (average 46 years). As average age for sphenoccipital chordoma is only 35, the author believes this supports the view already arrived at from the comparative frequency of the two conditions that only a small proportion of the cases develop into chordoma. They further state that ecchordosis physaliphora sphenoccipitalis if carefully looked for, is found to be by no means an infrequent occurrence although chordoma is a very rare disease. But rarity of cases of ecchordosis is obviously no index of their frequency. Stewart and Morin (1922) observed in lumbar region of fetus protrusions of noto-chordal tissue through body of vertebrae to both anterior and posterior surfaces. They suggested that the heterotopic tissue may be the starting point of neoplasia and the topographical distribution of chordoma certainly supports this view (rather than origin in nuclei pulposi of intervertebral discs themselves).

## 3. Site of Occurrence (chordoma)

68 cases described by Coenen showed distribution as follows:

### I. Cranial chordoma

#### 1. Clivus chordoma

a. Benign, 21

b. Malignant, 14

2. Hypophyseal, 1

3. Nasopharyngeal, 5.

4. Dental, 1

### II. Vertebral chordoma, 1

### III. Caudal or sacral

1. Antesacral, 13

2. Retrosacral, 8.

3. Central sacral, 4.

### Region of Sphenoccipital synchondrosis 25

(55 chordomas studied by Stewart and Morin):

1. Projecting into cranium, 16

2. Projecting into nasopharynx, 1

3. Projecting in both directions, 8

4. Occipital regions, 1

5. Upper and lower jaw, 1

Sacro-coccygeal region, 27

1. Projecting anteriorly, 12
2. Projecting posteriorly, 8
3. Projecting in both directions, 7
4. In lumbar region, 1

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Total -- 55

Chordomas other than in the occipital and sacral regions were thought rare by earlier observers. Coenen records only 1 case.

1928, Cappell (Glasgow) describes 3 new cases of chordoma of vertebral column. He believes that Trelet and Ranvier (1868) deserve credit for describing first case of cervical chordoma. Previously Syme and Cappell (1926) reported an interesting case in which the growth occupied a very unusual position, (cervical spine). At that time only 1 similar case in this situation had been recorded (Fabricius Muller 1918) but Klebs (1889) mentions another which perhaps belongs in this category. Cappell examined a collection of embryos in the Department of Embryology of Glasgow University. In the lumbar region of a 21 mm. embryo, small strands of notochord cells were seen extending from the central core which connects the notochordal masses in the intervertebral discs. As rule these strands passed in a ventral or lateral direction (occasionally dorsally), but it could not be demonstrated that these strands led to foci of chordal cells on the exterior of the vertebrae. In the sacrococcygeal region of this same embryo, small islets of notochord cells were seen on the anterior surface of the developing cartilage. Similar extensions of chordal tissue could not be demonstrated in the thoracic or cervical regions. These observations tend to confirm those of Berard Dunet and Peyron. Linck and Warstadt examined several embryos (30 to 50 mm.) and stated similar notochordal protrusions could not be found. Since the notochord, at an early stage, passes through all the vertebral bodies and connects the masses of chordal tissue (which eventually form nuclei pulposi,) there exists normally, apart from the presence of embryological remnants in abnormal situations, a basis from which notochordal tumors in vertebral bodies might arise. The factor injury in determining the onset of these growths seems to be clear in certain of the sacrococcygeal cases, and Ribbert's experiments

(1894) are also suggestive in this connection. No history of injury was elicited in any of Cappell's 3 cases.

Hutton and Young report 2 cases of malignant sacrococcygeal chordoma and 1 in the dorsal spine (1929). Davison and Weil report a lumbar chordoma (1928).

Note: Vertebral chordomas can occur and should be considered in vertebral body tumors.

#### 4. Pathology (Gross)

The gross appearance of chordoma varies with malignancy of growth; (degree of mucoid degeneration present is an index of comparative malignancy). In the typical case the tumor is well encapsulated and is broken up into lobules by fibrous trabeculae. Each lobule is composed of semi-transparent, whitish or bluish, gelatinous tissue, with or without a central focus of old or recent hemorrhage. The tumors to which it bears the closest gross resemblance are the more myxomatous types of "mixed salivary gland tumors, or colloid carcinoma". Chordoma in its later stages may become extremely gelatinous with breaking down of the interlobular septa. With increasing malignancy the tumor becomes more solid and opaque, the formation of mucin being in inverse ratio to the rate of cellular multiplication. One of the most striking features of chordoma, whether spheno-occipital or sacrococcygeal, is its locally destructive effect on bone. The base of the skull is penetrated and destroyed, often with extensions into nasopharynx, orbits or sinuses, and in the sacrum, coccyx and ossa innominata. Widespread infiltration is only seen in rare instances, and metastasis, so far confined to sacrococcygeal cases, is even less frequent. In the case reported by Stewart (1922); a large metastatic mass made its appearance over the right scapula. Regional lymph gland deposits are described by Peters and Pototschnig and cervical gland metastasis by Lewis. In Pototschnig's case there was also liver metastasis, and in Lewis' second case peritoneal and lymph gland dissemination occurred. Mathias' case of spheno-occipital tumor, not only involved orbits and sinuses but also eroded frontal bone, with fungation on surface.

In sacrococcygeal region growth may ulcerate into rectum (Debernardi, Albert)

but the skin retains its integrity in a remarkable manner even over very large tumors. Dunet and Peyron saw the largest chordomas so far reported, the former measuring 30 x 20 cm., the latter 26 x 22 x 15 cm. in diameter, each measuring 80 cm. in circumference.

#### 5. Microscopic changes

The notochord, of epithelial origin, develops along a connective tissue line into a structure having purely mechanical functions. So we find that while many chordomas are definitely epithelial in character, at least in part, others or portions of them are of the connective tissue type and when highly malignant, frankly sarcomatous in aspect. (Our first case). In spite of this diversity of structure, most chordomas present a highly characteristic appearance under the microscope. The main features are (1) alveolar character of growth, (2) solid epithelial aspect of younger, more cellular areas, (3) cytoplasmic and intercellular vacuolation, which is not marked in younger cells, but later becomes extreme, producing either so-called physaliphorous or bladder-like cells or where demarcation of individual cells is lost, the appearance of a high vacuolated syncytium, (4) formation, intracellularly, of mucinous fluid, escaping from cells, flows together to form first intercellular columns and later a sea of mucin in which only scattered cellular islets remain.

#### 6. Clinical aspects of chordoma.

Chordoma is usually tumor of low malignancy, slowly infiltrative and destructive, tending to recur after removal; exceptionally it grows rapidly, or give rise to metastases. The diagnosis can only be made with certainty by microscopic examination, but when a tumor occurs in the sacrococcygeal region it is often possible to express a clinical opinion with a fair measure of confidence as to its correctness.

Average Age in which the patient comes under observation was found to vary in the two groups. In 20 sphenoccipital cases it was 35 years; in 28 sacrococcygeal cases 50 years. The youngest and oldest patients in sphenoccipital series (16 and 72) in the sacrococcygeal series (22 and 68).

Sex Incidence is as shown in the following table:

|        | Spheno-<br>occipital<br>cases | Sacro-<br>coccygeal<br>cases | Total |
|--------|-------------------------------|------------------------------|-------|
| Male   | 13                            | 23                           | 36    |
| Female | 9                             | 6                            | 15    |

Note: More frequent in males.

Possible importance of trauma in etiology of sacrococcygeal chordoma is mentioned by several writers, (Berard, Dunet and Peyron - 1922). Severe coccygeal trauma might be a factor in liberation of chordal tissue from its normal osseous control, and even an incitement to proliferation. (Note Ribbert's experimentation on rabbits).

#### Chordoma of clivus

Clinical manifestations are those of rather slowly growing tumor involving base of skull and brain. Headache of varying severity, giddiness, nausea and vomiting, and progressive failure of vision are early symptoms noted and are followed or accompanied by palsies of various kinds depending on exact site and size of the tumor. Papilloedema may be present. Various combinations of cranial nerves may be affected, (5th and 7th most frequently) while in a certain number of cases the symptoms even from beginning are those of bulbar paralysis. Spastic hemiplegia or paraplegia may develop. In spite of the not infrequent involvement of the sella turcica it is rare to find evidence of pituitary disorder. An exceptional instance is seen in Lemke's case where there was polydipsia, and in which at postmortem, the pituitary was found to be infiltrated. X-ray examination affords valuable aid to diagnosis. In most cases the tumor grows upwards into cranial cavity, in others it extends both upwards and downwards. In 9 of 25 clivus cases it actually grew into nasopharynx and in one of these (Fabricius Muller's) there was no evidence of intracranial involvement. Projection of chordoma into the nasopharynx may lead to nasal obstruction or deafness.

Prognosis of sphenoccipital chordoma is very poor. Untreated cases run

average course of 2.8 years from first appearance of symptoms. Surgical interference has met with certain measure of success. In 1 case tumor was curetted from the base of the skull, but in spite of subsequent X-ray treatment patient returned with recurrence 7 months later. Fabricius-Muller's case, in which there was no evidence of intracranial involvement, was also treated by extirpation of growth. Recurrence occurred in few months and second radical operation was performed 4 1/2 years after first. Patient well 5 months later. In Spiess' case removal of the tumor by nasal route was followed by disappearance of all symptoms. In Hellmann's case the tumor was successfully extirpated. (No report of follow up).

#### Sacrococcygeal chordoma.

Pain, more or less severe, is earliest and most constant symptom. In early stages it is usually situated in sacral or coccygeal region; later there may be shooting pains in legs and buttocks. In later stages pressure on nerves may lead to paralytic and trophic phenomena in lower limbs and to interference with functions of micturition and defecation. Loss of touch, pain and temperature sense over penis, scrotum, and anoperineal region was present in Linck and Warstadt's case, dysuria, frequency, and later incontinence were found in many. Obstinate constipation is also a frequent symptom. The later symptomatology depends to a considerable extent upon whether the tumor is growing backwards, mainly forwards, or in both directions. Backward extension leads sooner or later to appearance externally of a tumor in sacrococcygeal region. It is median in position, slowly growing, firm, elastic, and bossed on the surface, and the skin moves freely over it. There is progressive destruction of sacrum and coccyx. Forward extension leads to appearance of a tumor in the hollow of sacrum with pressure on rectum, and in a few cases infiltration of wall. In 1 case the tumor had actually ulcerated through the rectal wall, with passage of blood and mucus.

Prognosis of sacrococcygeal chordoma is poor, although much better than in spheno-occipital cases. Average duration of symptoms in 18 cases (Stewart) in which death or recurrence is known to have taken place is 7 years, as compared with 3 years in clivus cases. Five cases were inoperable when first seen; in 3 of these an

exploratory operation was done; in a fourth, death followed partial removal of growth. In 21 cases the tumor was more or less completely excised. Three of these died soon after operation and in 3 no after-history was given. There remain 15 cases which survived operation, of whom no less than 13 developed recurrence; 4 died of recurrence, 3 were inoperable, and in 6 a second excision was performed. Of this last group 2 were not traced and 4 recurred a second time, 2 being dead and 2 inoperable at the time of writing (1926). In 2 cases only was the patient known to be alive and well at a date subsequent to the time of discharge from hospital. In one case reported five years elapsed before the recurrence was observed. In spite of these bad results, surgical extirpation should be attempted wherever possible on account of the long freedom from recurrence which is sometimes obtained. The results of the removal of recurrent tumors are less satisfactory. There is some amelioration of symptoms by partial excision.

#### IMPRESSIONS:

1. Benign notochordal rests were found in clivus by Virchow (1846).
2. Ribbert named the tumor "chordoma" in 1894, and reproduced it in rabbits by puncturing the intervertebral discs.
3. Tumor is probably not as rare as reported.
4. There are more than 80 cases now on record.
5. Benign rests outside bone of skull are found in 2 to 3% of all autopsies. They do not cause clinical symptoms.
6. Either heterotopic notochordal tissue or escape of confined tissue is the starting point of chordoma.?
7. Tumors usually occur in base of skull or sacro-coccygeal region.
8. Tumors of the other vertebrae are now recorded.
9. Tumors of the skull may extend down into nasopharynx, cranium, occipital regions and jaw.
10. They develop or extend in front of, in or behind sacrum.
11. Injury may be a factor in their development.
12. Gross appearance is mucinous, hemorrhagic, soft or solid.
13. They are locally destructive to bone, metastases is uncommon, but widespread infiltration may occur.

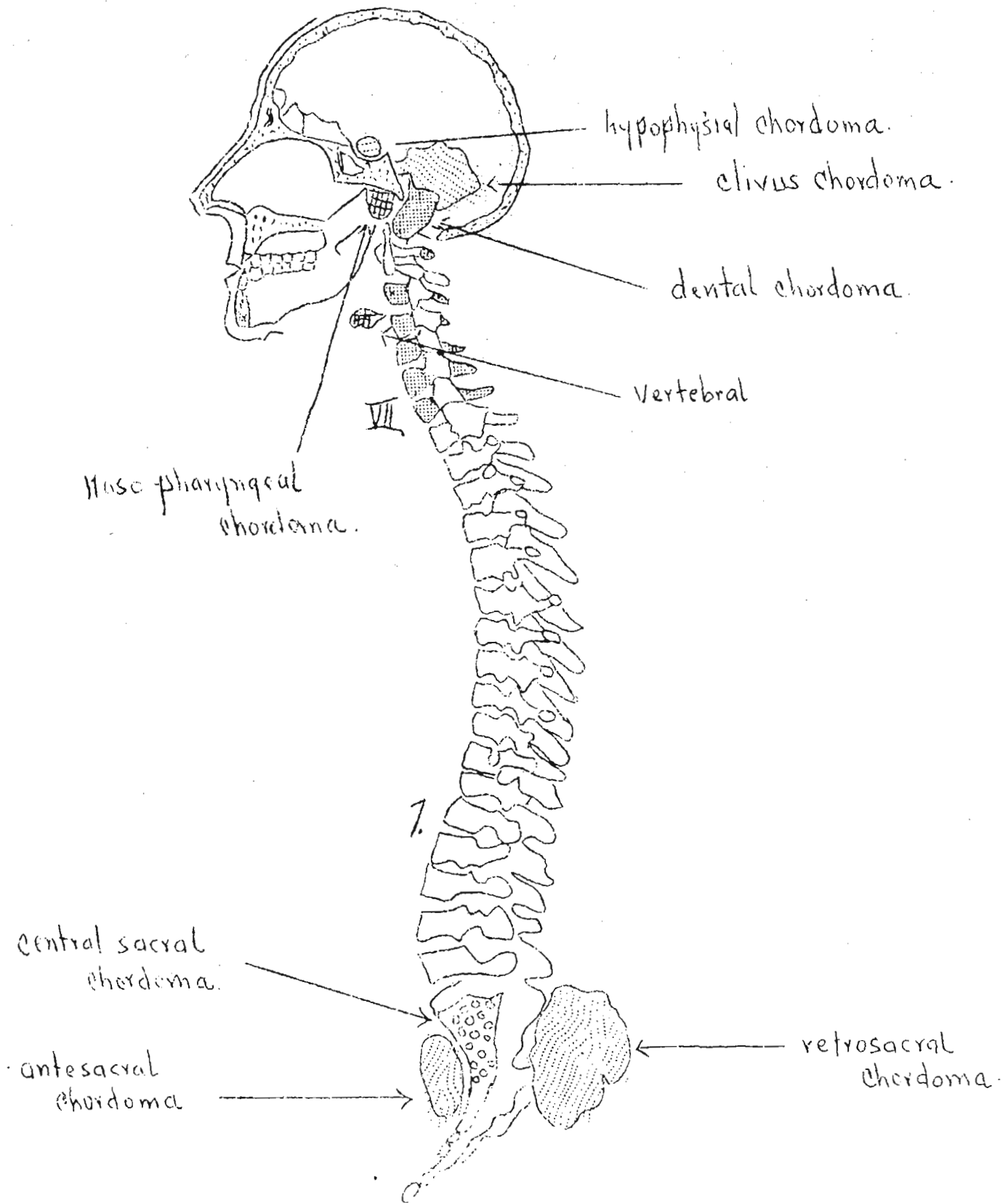


14. Skin is usually unbroken but may ulcerate from too heavy deep therapy ?
15. The microscopic structure is characteristic.
16. Tumor tends to recur after removal (usually low malignancy).
17. Average age of group in skull is 35 years, sacrum 50 years.
18. Males predominate.
19. Clivus chordomas give usual signs of brain tumor plus basal skull infiltrations.
20. Course of untreated skull cases is 3 years, sacrum 7 years.
21. Pain, soreness, sciatic syndrome, caudal involvement and obstipation are characteristic of sacral group.
22. X-ray finding of mid-line (or nearly so) solitary osteoclastic tumor of sacrum suggests chordoma.
23. Excision or deep therapy may be used.
24. Tumors tend to recur locally and prognosis for life is usually not good.

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Note: The last reference is such a complete exposition of our knowledge of the intervertebral disk that it was not abstracted. The entire article should be read by all interested in this subject.



Schema of distribution in chordomas  
clinical and anatomical types.