Hypertrophic Pulmonary Osteo-Arthropathy

As a final word I would venture to recommend your especial attention to the following points:

1. The dosage of urotropin should be increased until an antiseptic action is obtained or the limits of tolerance are reached.

2. The reaction of the urine should be carefully tested and the acidity if necessary raised by means of drugs until the point of dissociation of urotropin is reached.

3. Diuretics and diuretic waters interfere with the splitting of urotropin and should be avoided during its administration.

4. The Rimini-Burnam test for formaldehyde should be in constant use when urotropin is being administered.

5. Urinary antiseptics of the formaldehyde series are harmful or useless: (a) In very acute inflammation of the urinary organs; (b) in pure tuberculous infection of the urinary organs; (c) when the urine is alkaline.

Reference.—1 Lancet, December 1900.

Hypertrophic Pulmonary Osteo-Arthropathy
(Marie's Disease).

By A. Gilmour, M.D., M.R.C.P.E., D.P.H.,
School Medical Officer, Paisley.

At the medical inspection of scholars attending a large special school for physically and mentally defective children, I was interested to discover a case of hypertrophic pulmonary osteoarthropathy. As it is rare to find records of this disease, described by Marie, occurring in so young a patient, a few particulars may be of general interest. Many details about the case could not be investigated, as the greater part of the examination had to be made during school hours, and the boy, being of a peculiar temperament, resented a thorough examination.

The history was as follows:—

X. Y., aged 9 years. The boy lived with his grandfather, under whose care he came when about 1 year old. The mother died of what was thought to be "consumption," while the father, who is alive, is stated to be "unsteady" in habits. No further history of tubercle could be obtained, and there was no specific history. One other younger child is stated to be healthy. The social conditions and surroundings are good.

The early history of the boy is best given in extracts taken from the records of the Royal Alexandra Infirmary, Paisley, in which institution he was treated on three occasions.

At the age of 16 months he was admitted "with a sinus over the terminal phalanx of the fourth toe of the left foot. Probe reached
bone. The toe is greatly swollen, and the first phalanx is also thickened. Toe amputated at the metatarsal-phalangeal joint. Patient dismissed 'well' in two days."

On the 28th September 1906 (aged 4½ years) he was admitted "with tubercular right knee-joint. The tissues were swollen, and there was a little fluid in the joint, but no evidence of disease of the cartilages. The limb was fixed in a posterior splint, and a Scott's dressing applied. Dismissed on 9th November 1906 cured."

On the 21st March 1907 he was again admitted "with swelling of right knee-joint; large and more or less painless; the natural hollows obliterated, but not much muscular wasting. There is considerable thickening of the tissues round the knee-joint. Little or no fluctuation. Knee excised 28th May 1907, and the boy was dismissed on 29th June 1907 cured."

Shortly after this the spinal column was noticed by the guardians to be becoming curved and a plaster-of-Paris jacket was applied. This did little good and was soon discarded.

Subsequent History.—Two years ago the fingers and wrists were noticed to be "swollen," and a little later the toes and ankles were similarly affected. About the same time he suffered from some "breathless attacks," but he never seems to have been fevered at nights, and has never had night sweats or diarrhoea. The conditions all appear to have been practically painless.

Present Condition.—Weight 51½ lbs. (3 months previously 53 lbs.). The boy is small and very emaciated, and the kyphosis of the spine and the enlargement of the wrists and hands give him a peculiar ape-like attitude. The face is small, but well formed, the bones of the face and cranium being unaffected. The complexion is clear and slightly waxy, with a faint cyanotic tinge in the hectic flush on each cheek. The general muscular development is poor, and there is no subcutaneous fat, so that the bones are all easily palpable.

As the arms are symmetrical in appearance and measurement, one description covers both. The humerus is unaffected. About the lower third of the forearm the radius and ulna begin to thicken, increasing gradually towards the lower ends of the bones. The condition is hard and bony. The wrist joints appear "slacker," and the lower margins of the radius are felt to be distinctly thickened.

The metacarpal bones are slightly enlarged, broadening the hands. The proximal phalanges seem "baggy" in their centres, giving a thickened appearance. The middle phalanges are less affected. The terminal phalanges are markedly "clubbed," but the colour of the hands, wrists, and fingers is pale and waxy. On palpation of the terminal phalanges the enlargement is fairly firm, but appears to be more confined to the soft tissues, and there is some flattening anteroposteriorly. The nails of the fingers and thumbs are much enlarged, curved both vertically and horizontally. They are smooth, fine, of a
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Fig. 1.—Shows clubbing of fingers and overgrowth of nails.

Fig. 2.—Hand, left. Shows marked enlargement of distal ends of terminal phalanges; general enlargement of the long bones of hand, especially of metacarpal of little finger, with deposit of lime salts and new bone; periosteal round shaft; carpus well marked. A very slight absorption of lime salts from all bones. Epiphyses well marked. General prominence of soft parts.

Fig. 3.—Forearm, left. Periosteal deposit of lime, well marked, and irregularity of bony cortex like chronic osteomyelitis. Whole of both bones expanded and thickened, irregular lacuna in cortical bone, medullary canal practically obliterated.

Photograph and Skiagrams by Dr. Hope Fowler.
Hypertrophic Pulmonary Osteo-Arthropathy 529

yellow-pink colour, and owing to the curvature come well over the ends of the phalanges. There is no evidence of congestion, and the hands are fairly warm.

The lower extremities exhibit similar appearances. The hip-joints and thighs are unaffected. The right knee has been excised and is firmly ankylosed by bony union. The curved anterior cicatrix of the operation is white and firm. About the lower third of the leg the tibia and fibula begin to thicken and enlarge. The ankle-joints are free in action. The feet appear thickened and swollen over the instep, and the phalanges present an appearance similar to that of the hands, the fourth left digit being wanting (see history). The nails are much enlarged and similar in character to those of the hands.

The measurements of both the upper and lower extremities of the patient were symmetrical.

The following measurements of the upper and lower extremities are contrasted with those obtained in the case of a boy of about the same physical development, suffering from congenital heart disease and exhibiting marked "clubbing," and with the average of similar measurements in twenty boys who were of the Anthropometric Committee's standard for the same age. As this standard is high, and as a comparison would more scientifically be made with boys of the same physical development, it follows that the difference is the more striking.

**Measurements in Inches.**

|                         | Patient. | Cardiac. | Standard. |
|-------------------------|----------|----------|-----------|
| **Upper Extremity—**    |          |          |           |
| Circumference of upper arm | 5.0     | 5.0      | 6.5       |
| " of upper end of forearm | 6.25    | 6.0      | 6.93      |
| " of middle forearm     | 5.37    | 5.25     | 6.07      |
| " immediately above wrist joint | 6.0  | 4.0      | 4.87      |
| Length of radius         | 7.5     | 6.5      | 7.12      |
| Wrist joint to tip of middle finger | 6.25  | 4.8      | 5.18      |
| Circumference of proximal phalanx of middle finger | 2.12   | 1.5      | 1.96      |
| " of second phalanx of middle finger | 1.12   | 1.4      | 1.75      |
| " of terminal phalanx of middle finger | 2.0    | 1.35     | 1.54      |
| " of proximal phalanx of thumb. | 2.12   | 1.62     | 2.0       |
| " of terminal phalanx of thumb. | 2.5    | 1.6      | 1.8       |
| Breadth of thumb nail    | 0.94    | 0.62     | 0.56      |
| **Lower Extremity—**     |          |          |           |
| Circumference of thigh   | 9.0     | 10.0     | 13.5      |
| " of calf                | 7.5     | 6.75     | 9.5       |
| " of lower end of tibia and fibula | 8.0  | 5.25     | 6.5       |
| " of terminal phalanx of first toe | 3.12 | 2.5      | 2.75      |
| " of terminal phalanx of middle toe | 2.0  | 1.5      | 1.6       |
| Breadth of nail of first toe | 1.0    | 0.75     | 0.64      |
The spine exhibits marked kyphosis beginning in the mid-dorsal region, the upper dorsal vertebrae being curved forward almost at right angles to the lower. There is no scoliosis, and both sides appear symmetrical. As a result of the kyphosis the head appears to lie sunk between the scapulae and clavicles, the lumbar curve is lost, and the sacral spines appear more prominent. There is no tenderness over the spine, but the boy resents examination.

The capacity of the chest is altered by the kyphosis, being almost circular in outline at the nipple line. The upper part is narrow, and the lower ribs meet the sternum at an acute angle. The sternum is flat, narrow and pointed. Examination of the respiratory system reveals no cough, no spit; increased vocal fremitus and resonance all over, with well-marked bronchial breathing, with occasional dry râles on the right side; percussion is dull all over. The heart is slightly enlarged with an accentuated second sound.

The abdomen is prominent and tympanitic all over. The edges of the liver and spleen are both palpable. The urine is dark amber; no abnormal constituents. There are no signs of peripheral neuritis or other nervous phenomena. Handling of the limbs, however, makes the patient cry out, as "he does not like them touched." He is extremely childish and emotional.

Examination, although gentle and apparently painless, is resented, and the boy cries for no apparent reason. He is bright and intelligent. He can walk fairly well but is easily fatigued. He suffers no pain. Sleep is usually undisturbed.

Subsequent History.—The patient was under observation for five months, during which time no change in the physical condition occurred with the exception of slight loss in weight. He complained at times of "feeling cold," but appeared to suffer no pain.

About the end of June the boy was reported "absent" from school, but no indication of the nature of the illness was given. He died six days later from an illness with all the signs of tubercular meningitis. No post-mortem was granted.

The diagnosis of this case is simple, the history and features being typical of hypertrophic pulmonary osteo-arthropathy. The disease is distinguished from acromegaly by the non-involvement of the face and cranium, by the symmetrical enlargement of the bones of the wrists, ankles, etc.

The case is of interest, more especially from the age of the child. Though the disease is most common between the ages of 20-25 years, records show that it may occur at any age. Davis has described a case in a child aged 4½ years, suffering from empyema communicating with a bronchus. Bamberger mentions a typical case at 7 years, and Whitman one at 8 years. Other
cases have been described in young children, but investigation shows that these are usually cases of "clubbing" without any bony changes. The present case appears to have been characteristic at 7 years of age.

Since the disease was originally described by Marie, many observers have recorded cases and theorised on the causation, but an explanation applicable to all has yet to be found. Marie was of the opinion that the condition was always secondary to some chronic lung condition in which toxins were produced which had an "elective" action on certain parts of the osseous and articular systems producing inflammatory changes. Bamberger described similar changes in "chronic pulmonary and cardiac diseases," but held that the lung cases were all "bronchiectatic" and nontubercular—the essential point being the presence of a cavity from which "chemical substances" were absorbed which gave rise to the osseous and articular inflammations.

Thorburn, describing three cases and analysing previous records, was of opinion that the condition was "a tubercular affection of a large number of bones and joints, but that it is of a benign type, having no tendency to break down or caseate." He attributed the location of the disease to the feebleness of the circulation, and suggested that the disease should be called "tuberculous polyarthritis." In the report of a post-mortem examination he concludes:—"The periosteal inflammation is one which the tubercular virus is competent to produce, and closely resembles the periostitis often found on the shafts of bones affected by tubercular epiphysitis. The change in the synovial membranes of the knee was also of the type of tubercular disease, but the other joints were further removed from this type, and all that can be said of them is that they suggest the action of a chemical poison rather than of an organised irritant."

In a thesis on this disease Alexander analysed 77 typical cases, in which the excess of males (64) over females (13) is clearly demonstrated. In these cases bronchiectasis was found in 25 cases, chronic pulmonary tuberculosis in 15, empyema in 8, and malignant diseases of the thoracic organs in 8. The disease, however, also occurred in cardiac, hepatic, and biliary conditions, and in two cases no cause could be discovered.

The synopsis of post-mortem appearance of 16 cases in which the bony changes were investigated shows a "symmetrical deposit of new subperiosteal bone," most common at the lower ends of the bones of the forearm, in the metacarpals, and the first two rows
of phalanges. The bones of the lower extremity were less frequently affected, while the clavicles, ribs, and iliac crests were occasionally involved. The periosteum was thick and vascular, the nutrient arteries were enlarged, and additional blood-vessels were present. No change has been mentioned in the cancellous tissue, and only one case showed softening of the compact bone. The synovial membranes of the joints showed inflammatory changes, but the articular surfaces were unaffected except in two cases in which slight erosion was described. The clubbing of the fingers is due to a hyperplasia of the soft parts, both fatty and fibrous. There were no changes in the bone of the terminal phalanges.

The formation of spicules of bone at the terminal phalanges has been described by Hall, but such changes are said to occur as frequently in healthy subjects. Hypertrophic pulmonary osteo-arthritis therefore would appear to be most frequently found associated with diseases in which there is pus formation, or breaking down of tissues, with retention of the secretions. Syphilis appears to be an unimportant factor, while nerve conditions, central or peripheral, appear coincidences. Möbius describes a case in which clubbing of the fingers occurred in a neuritis of the ulnar nerve.

In Allbutt and Rolleston's *System of Medicine* the disease is spoken of as "Marie's sign-group," in which the affection is considered to be "a group of morphological conditions, consisting (1) of clubbing of the fingers and toes, to which none of the various synonyms draw attention; and (2) of certain changes in the ends of the long bones, more especially in the wrist and ankle-joints, and of the cartilages and synovial membranes in these and occasionally in other joints."

The evidence is discussed under these two headings, the "clubbing" of hypertrophic pulmonary osteo-arthritis being stated to be similar to clubbing in other conditions.

From the statements given above, it would appear as if the condition was produced by a chronic toxæmia, usually bacterial, but occasionally that of altered body metabolism. The improvement in the condition, resulting from removal of the toxic cause, would support this theory. It is possible that the condition may be one in which the amount of calcium salts in the blood plays a definite part in the overgrowth of bone formation. In the present case the examination of the blood was unfortunately omitted. The difficulty is to explain those cases in which a
unilateral enlargement has been described. The more rapidly the disease develops, the more probable appears to be the involvement of the joints.

Other names, such as "secondary hypertrophic osteo-arthritis" by Wynn, have been suggested for the condition, but probably the new designation, "Marie's sign-group," is least erroneous.

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THE ROYAL COLLEGE OF SURGEONS OF EDINBURGH: THEIR PROFESSORS OF SURGERY.

By C. H. CRESWELL, F.S.A.(Scot.).

Although Mr. James Russell was the first Professor of Clinical Surgery, the idea of clinical teaching did not originate with him. The credit of applying this method of study to surgery perhaps belongs to Mr. James Rae, who also had some reputation as a dentist. Mr. Rae appears to have taught surgery some years previous to 1772, for in that year, in reply to an application from him, the College agreed to recognise his lectures, and ordered the following notice to be inserted in the public press:—

"The College of Surgeons being desirous to promote every usefull undertaking towards the advancement of the knowledge of Surgery, have taken into their Consideration A PLAN of Lectures on the whole Art of Surgery; Also practical discourses on the Cases of Importance as they occurr in the Royal Infirmary, given for several years past at their Hall by James Rae, Surgeon in Edinburgh and one of the members of the Society. As this course is founded on the practice of the Hospital and delivered by a person who has been in the habit of Constant Observation, they recommend it as usefull and necessary to the Students of Physic and Surgery, and to render this course more extensively usefull the Society are resolved to Communicate to him such cases of Importance as may occur in their practice."

Mr. Rae's endeavours seem to have been successful, and four years later he again applied to the College for assistance. This