

Additional experience with this new method is necessary, but even if the present good correction is lost or a new deformity develops the child can be treated by external splintage followed by a triple arthrodesis at the age of 12. It is intended to show her again in five years' time.

An Unusually Large Foreign Body in the Hand.—F. G. ST. CLAIR STRANGE, F.R.C.S.

A FARM labourer had his right hand impaled by a Rototiller blade, and was brought to hospital with the foreign body *in situ*. [The blade (Fig. 1), which measured $4\frac{3}{8}$ in. by 5 in. overall and weighed $9\frac{3}{4}$ oz. had entered the ulnar border of the wrist and emerged through the palmar aspect of the thumb. Owing to its curvature, removal was impossible without open operation, when it was found to be lying between the tendons and nerves in front of the wrist and within the abductor pollicis brevis.]

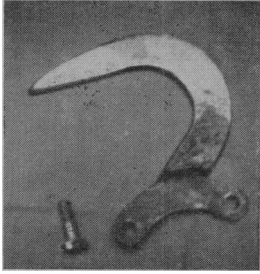


FIG. 1.—The rototiller blade, measuring $4\frac{3}{8}$ in. \times 5 in. The width of the blade is up to $\frac{7}{8}$ in. and its thickness $\frac{1}{4}$ in.

Post-operatively, there was temporary median palsy and anaesthesia, but this recovered, and he was left with an almost normally functioning hand, the only residual disability being incomplete flexion of 5th finger.

A general discussion on the case showed that the majority of members were in favour of postponing any attempt to free the flexor tendons to this finger, as further improvement by resolution was to be anticipated.

Progressive Familial Hypertrophic Polyneuritis (Dejerine-Sottas Syndrome, 1893).—MICHAEL J. COX, M.B.

Miss B. W., aged 55.

She was born with normal feet except for a high instep but she always had a deformed back.

Aged 7 her family noticed she would trip over easily and she began to turn her feet inwards and then between the ages of 11 and 14 she had special exercises for her feet.

Aged 14 she underwent an operation for bilateral pes cavus. She then walked fairly well until the age of 30 when "a sudden shock took her off her feet". Since then she has crawled most of the time on her knees, occasionally pulling herself up to stand on the dorsum of her feet.

Aged 45 onwards she began to develop ulcers on both feet which have become very painful in the last few months.

Her right hand is deformed but has a good grip while there is good function in the left hand.

Social history.—She has been caring for her mother for many years doing quite a lot of housework but no cooking. She has not been in a shop for about twenty years and she has not been out of the house for five years.

Family history.—Her grandfather suffered from "curvature of the spine" and was called "high-kicker" because of an abnormality of his feet. Her father suffered from a minor abnormality of his feet; one of her aunts had talipes equino-varus and absent deep reflexes, while another aunt suffered from curvature of the spine. One of her cousins suffered from "club feet" and two other cousins suffered from the fully-developed syndrome similar to this patient. Two of her nieces suffered with "curvature of spine", one associated with pes cavus, and one has absent deep reflexes.

This family has been previously reported by Russell and Garland (1930).

Physical examination.—Cranial nerves normal. Gross kyphoscoliosis convex to right in the thoracic region and to the left in the lumbar region.

Upper limbs: There was wasting of the arms and severe wasting of the intrinsic muscles of the hand, right worse than left. There were flexion contractures at the proximal interphalangeal joints of all fingers of the right hand and of the ring and little fingers of the left hand. Ulnar nerves thickened, right larger than left. Reflexes absent. No sensory disturbance.

Abdominal reflexes absent.

Lower limbs: wasting of thighs and calves. Thickened callosities over both tibial tubercles and thickened infrapatellar bursae. Power good in both hips and knees. Both lateral popliteal nerves were greatly thickened.

Feet (Figs. 1 and 2): Gross bilateral talipes equino-varus, A large penetrating ulcer under the right heel and a smaller one on the left. The reflexes were absent but there was no sensory disturbance.

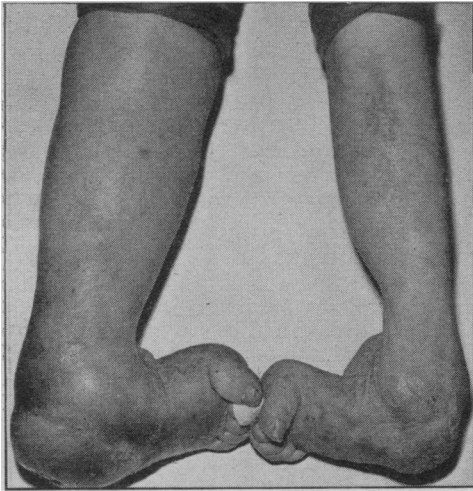


FIG. 1.

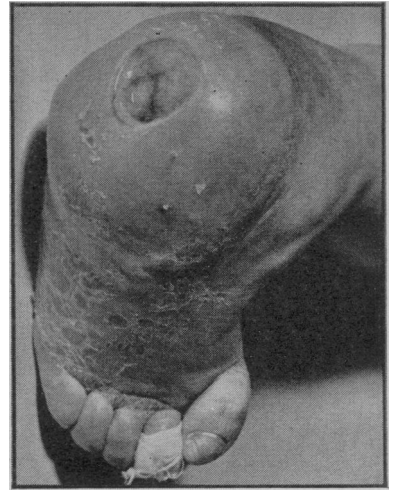


FIG. 2.

Progress.—The ulcers were cleared up with local treatment and systemic chemotherapy. Then a left Syme's amputation was performed by Mr. H. Osmond-Clarke. The os calcis was found to be very mushy and was removed piecemeal. A right Syme's amputation was performed on 15.10.55.

Pathology.—Macroscopic: Dissection of the amputated left foot showed extensive fatty replacement of the flexor digitorum brevis and other muscles. The cartilage of the talus was greatly eroded by osteoarthritis. The nerves were diffusely thickened (medial plantar, 0.5 cm. in diameter; lateral plantar 0.35 cm. in diameter at points close to their origins). There was less thickening of the anterior tibial, musculo-cutaneous and sural nerves.

Microscopic: (1) *Nerves.* The five nerves mentioned were examined in longitudinal and transverse sections. The thickening is mainly due to a great increase of collagen throughout the epi-, peri- and endoneurium. In the first two layers it is dense and hyaline; in the last mentioned, fibrillar. There is also proliferation of the endoneurial cells and, occasionally, of similar cells in the perineurium. In the endoneurium the cells tend to form onion-like whorls; the individual cells are separated by delicate collagen fibres. These whorls are formed about axis-cylinders, but a large proportion of the latter have undergone destruction. There is no inflammatory cellular reaction other than a rare and small group of small lymphocytes. The blood vessels are not significantly altered.

(2) *Muscles.* In these there is gross replacement by adipose tissue. Such muscles as persist are almost all greatly atrophied, though a few are hypertrophic. There is conspicuous collagenous thickening of the endomysium. Attempts to demonstrate motor nerve-endings by the methylene-blue technique were unsuccessful.

Discussion.—This case is presented to show another of the indications for the Syme's amputation. By means of this treatment it is hoped to be able to mobilize this woman who has led such an incapacitated life for years.

This disease bears close resemblance to Charcot-Marie-Tooth peroneal atrophy while the enlargement of the nerves is seen in Von Recklinghausen's disease.

The possibilities are discussed by Russell and Garland (1930) who thought it advisable to class it as a separate entity.

Summary.—A case of familial hypertrophic polyneuritis, treated by Syme's amputation, is presented.

I should like to express my thanks to Sir Reginald Watson-Jones for his encouragement in the presentation of these two cases and to Professor Dorothy Russell for her very great help in preparing the report of the pathology of this case.

REFERENCES

- DEJERINE, J., and SOTTAS, J. (1893) *C.R. Soc. Biol., Paris*, 5, 63.
 RUSSELL, W. R., and GARLAND, H. G. (1930) *Brain*, 53, 376.

[This Meeting will be continued]