that is, intermediate between the changes described by Marchesani and by Marfan. However, in the hands shortening of the digits, broadening of the metacarpals in one subject, and flexion deformities of the fingers due to capsular and tendon contractures are isolated brachymorphic features. These features are similar to those of Leri's pleonoteosis (Watson-Jones, 1949), in which flexion contractures of the interphalangeal joints are associated with broadening and shortening of the thumbs and great toes. In contrast, we sometimes found isolated dolichomorphic characteristics in the same hand such as ligament laxity and slenderness of the phalanges.

The constant finding of a high patella in the more severely affected members of the family is another feature of the dolichomorphic end of the spectrum. The expression of the defect appeared to be readily modified by other genes situated at the same locus. The mother of the propositus had flexion deformities of all the fingers except the little finger of the right hand, which was deformed by a swan-neck type of hyperextension deformity; yet in her daughter, the propositus, the swan-neck deformity affected all the fingers except the little finger, which was the site of a flexion deformity. It is possible that the selection of the joints involved in certain forms of arthritis may similarly be influenced by individual variations in the genetic structure of joints and tendons.

Summary

A musculoskeletal disorder resulting in flexion and swan-neck deformities of the fingers is described for the first time.

The disorder affected 10 female members of one family and was inherited as a Mendelian sex-linked dominant characteristic.

Excessive slenderness of the phalanges and shortening and broadening of the first and second metacarpals were observed in radiographs of the hands of certain members of the family.

The digital abnormality was found to be associated with the presence in the urine of an abnormal substance having the characteristics of an α-amino-acid in all affected members of the family, and also in two members who appeared to have normal fingers.

The relationship of the new syndrome to recognized heritable dolichomorphic and brachymorphic disorders of connective tissue is discussed.

We wish to thank Professor A. L. Latner for his help and encouragement in this work.

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Preliminary Communications

Use of Pronethalol in Phaeochrome Tumours

The introduction of effective alpha-adrenergic blocking drugs (phenoxybenzamine and phentolamine) has diminished the risk of excessive hypertension, and of most arrhythmias, before and during surgical treatment of phaeochromocytoma. These drugs do not, however, prevent excessive cardiac stimulation due to beta-activity of adrenaline and noradrenaline, which may produce ventricular tachycardia and fibrillation.

There were thus strong *a priori* grounds for the trial of pronethalol in the following cases in addition to the phenoxybenzamine.

Case Reports

Case 1.—A young woman was treated at St. Thomas's Hospital under Dr. Evan Jones. She had severe sustained hypertension and had had attacks of paroxysmal dyspnoea. The biochemical findings were characteristic. She was prepared for operation as usual with phenoxybenzamine, which mitigated the hypertension, but tachycardia of 130/min. persisted, as did pulsus alternans and a cardiac triple rhythm. A trial dose of 50 mg. of pronethalol was given intravenously. This slowed the heart rate to 90/min. and the pulsus alternans and triple rhythm disappeared. At the same time the jugular venous pressure rose, the superficial veins constricted, and the patient sweated. However, there was little drop in blood-pressure, and she felt no ill-effects. (In view of the reports by Stock and Dale (1963) of the precipitation of failure in stressed hearts, we now realize that intravenous administration should be avoided in such cases where possible.) The untoward effects soon passed, and the patient was maintained on oral pronethalol (400 mg./day) plus phenoxybenzamine, with a fall in pulse rate from 130 to 90/min., up to the operation. The operation was carried out, without any undue tachycardia or hypertension, by Mr. K. Shuttleworth, who removed the tumour. After initial improvement the hypertension returned and it became apparent that another tumour was present. This time the patient was prepared with the alpha- and beta-adrenergic blocking drugs orally, and a second tumour was removed with only a brief incident of a possibly nodal tachycardia during manipulation of the tumour.

Case 2.—A man aged 45 had had a large phaeochromocytoma removed from his right adrenal in 1955. He was readmitted to University College Hospital under Professor M. L. Rosenheim in March, 1963, because of a recurrence of his attacks. The attacks had recurred some years previously, when a laparotomy had not revealed a further phaeochromocytoma, and had been controlled by phenoxybenzamine. On this readmission his attacks recurred, with an excess of pressor amines in his urine, when the phenoxybenzamine was stopped. He was prepared for operation by the addition of 300 mg. of pronethalol a day for the two days prior to the operation and 100 mg. on the operation morning. He underwent a prolonged laparotomy, during which a small phaeochromocytoma was removed from behind the inferior vena cava, also some small remnants from the right adrenal bed. During the operation, by Mr. Bernard Harries, there was no undesirable tachycardia or hypertension.

Comment

In the light of experience of the first case and the findings of Stock and Dale (1963) in patients near heart failure, we would think it wise to avoid intravenous pronethalol where possible in these cases. Moreover, we would expect that its administration to patients with phaeochromocytoma *not* already receiving phenoxybenzamine might cause a serious rise in blood-pressure by abolishing the dilator (beta) action of adrenaline on peripheral vessels, especially

in muscle, while leaving the constrictor (alpha) actions unopposed.

We would suggest on this limited experience the following pre-operative regime: phenoxybenzamine 1 mg./kg./day orally, plus pronethalol 300–400 mg./day (in three or four doses), for an adult, both drugs for three days before surgery.

If, during operation, undue tachycardia or abnormal rhythms occur, intravenous pronethalol might be necessary —say, 25 mg. at first.

We are grateful to the clinicians in charge of these cases for leave to refer to them here.

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Medical Memoranda

Accidental Loss of Plastic Tube into Venous System

With the increasing use of the soft intravenous polythene catheter, an interesting complication is occurring with what appears to be disconcerting frequency. Moreover, this complication of the "lost tube" is associated with a surprisingly high morbidity and mortality, according to the collected statistics of Taylor and Rutherford (1963).

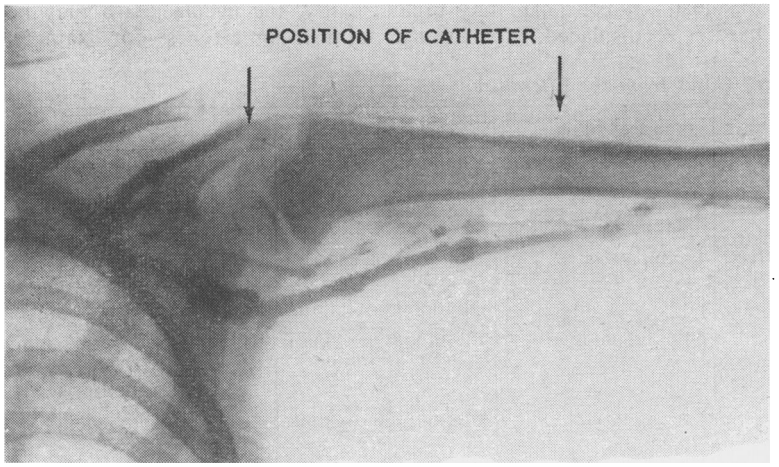

FIG. 1.—Venogram showing cephalic and axillary veins, with catheter (filled with dye) in the cephalic vein.

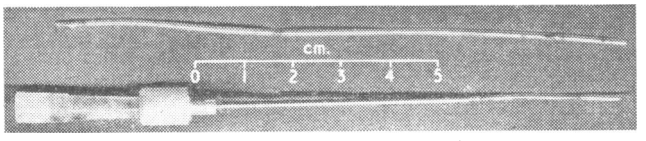

FIG. 2.—The two pieces of the catheter.

CASE REPORT

A 3-year-old girl had a skin-grafting operation for scalds of arm, chest, and neck. She was anaesthetized with thiopentone and D-tubocurarine, and then an intravenous drip was started through the left median antecubital vein just distal to the elbow, using a Bardic Deseret intracath. This type of apparatus is in common use in the Royal Liverpool Children's Hospital. It consists of a very sharp needle through which a soft polythene catheter 8 in. (20 cm.) long is introduced into the vein, the needle being then withdrawn.

Just before the end of the operation the drip was taken down, and it was then observed that only part of the catheter had been removed. A piece about 6 in. (15 cm.) long had sheared off during introduction, probably owing to inadvertent withdrawal of the catheter against the sharp needle-edge at some stage, and was lost in the child. Firm digital pressure was immediately exerted over the left axillary and cephalic veins. A plain *x*-ray film taken on the operation table did not reveal the presence of the catheter. The arm and axilla were palpated for the presence of the catheter. A firm "cord" was felt in the pectoral region, but on exploration through a small incision this proved to be a tendinous band of the pectoral insertion. The terminal part of the cephalic vein was identified and was seen to have a normal calibre and blood flow.

An injection of 6 ml. of a radio-opaque dye (30% sodium acetrizoate; "diaginol") was made into the cephalic vein at the wrist; firm digital pressure was maintained over the axilla apart from momentary release while the *x*-ray picture was being taken. The film clearly demonstrated the catheter in the cephalic vein (Fig. 1). The catheter was removed through a small appropriately sited incision (Fig. 2).

COMMENT

It is very commonly believed that this complication is rare, and that inert polythene tubing in the venous system is innocuous and does not merit determined efforts for its removal. That these beliefs are unfortunately not founded on facts is borne out by the reports of Moncrief (1958) and Taylor and Rutherford (1963).

Thus there was an incidence of lost catheter in 3 out of 135 cases as reported by Moncrief, stressing the need for very careful use of this invaluable but potentially dangerous mode of intravenous therapy. Moreover, of the 11 cases collected by Taylor and Rutherford, five patients died from infection as the direct result of the lost catheter, two died from causes unassociated with the intravascular catheter, and two underwent a right atriotomy for removal of the catheter. This experience of Taylor and Rutherford prompts them to suggest that every lost catheter should be removed, and that if for any reason a "waiting policy" is adopted an unexplained rise of temperature is a definite indication for right atrial exploration. Septic venous thrombosis is the commonest cause of death, though myocardial necrosis has also been described. The catheter usually ultimately lodges in the right atrium, impinging against the tricuspid valve, trailing back into the superior or inferior vena cava, but may occasionally negotiate the tricuspid valve and lodge in the pulmonary artery.

Should this complication occur we suggest that a contrast venogram may be of great value in locating the lost piece of catheter and facilitating its removal. A radio-opaque fibre or material incorporated in the catheter would also be advantageous.

We wish to thank Mr. P. P. Rickham for permission to publish this case, and Miss I. Forshall and Dr. G. J. Rees for constructive criticism. We also wish to thank Mrs. Kenworthy for help with the *x*-ray films and Miss Tankard for secretarial assistance.

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