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Pseudo-pseudo-hypoparathyroidism with coarctation of the aorta— a clinical syndrome?

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Summary

Two cases of pseudo-pseudo-hypoparathyroidism are described, associated with coarctation of the aorta, suggesting a further common clinical link (besides those already recognized) between the former condition and Turner's syndrome. The need for a change of nomenclature of pseudo-pseudo-hypoparathyroidism is emphasized.

Case no. 1.

A single woman, aged 50, was referred for opinion because of recent dyspnoea. She had not been allowed to play games at school because of 'some heart condition', but had led a normal life in all other respects, and had always menstruated normally. Her father had died of 'a heart complaint', her mother was alive but was being treated for pernicious anaemia, and her sister had 'rheumatism'.

On examination she was short (height 145 cm) and weighed 62 kg. She had a rounded facies, small hands and feet, and a recessed knuckle of the left

fifth finger. BP 240/100 mmHg in the right arm. No femoral or other leg pulses could be felt, but there was a prominent carotid pulse bilaterally and collateral arterial pulsations were felt over the thorax. No thrills were felt, but there was a loud, high-pitched ejection-type systolic murmur all over the front of the chest. A chest X-ray showed gross notching of several ribs, an elongated aortic knuckle, and post-stenotic dilatation of the descending aorta. X-ray of the hands confirmed their small appearance and the shortening of the left fifth metacarpal. An ECG showed left ventricular hypertrophy with negative T waves in leads 1, VL and V6. The patient refused admission to hospital for further investigation to confirm the clinical diagnosis of coarctation, and to exclude associated cardiac lesions; but it was felt that the diagnosis was reasonably certain in view of the physical signs and radiological features. The following investigations were normal: blood urea and electrolytes, routine urinalysis, serum calcium, phosphorus and alkaline phosphatase, haematological

profile, plasma protein electrophoresis, protein-bound iodine, diurnal variation of plasma cortisol, total urinary 24-hr excretion of 17-hydroxycorticosteroids, and chromosome examination. Her eye examination was normal, but she had very carious teeth. Her relatives were not accessible for examination and it seemed likely that they shared the patient's obviously subnormal mental state (which was not submitted to formal IQ assessment).

Case no. 2.

A single woman, aged 25, a machine operator, was referred for opinion because of exertional dyspnoea and oedema of the ankles for 2 years. There were no other cardiac symptoms, nor any such features in her past or family history. Her previous medical history was uneventful and menstruation normal.

On examination she was short (height 157 cm) and weighed 87 kg. Her mental ability was normal, but she had short hands and feet, and the knuckles of both fifth fingers were recessed. BP 130/70 mmHg. The right femoral pulse was very weak and the left was not felt; the popliteal pulses and ankle pulses could just be felt with difficulty. There was slight oedema of the ankles but no increase in jugular venous pressure. There was a moderately loud ejection-type systolic murmur at the mitral area and at the pulmonary area. The serum calcium, phosphorus and alkaline phosphatase were normal, as was a chromosome examination. A chest X-ray showed a prominent rounded aortic knuckle, with slight general dilatation of the aortic arch, and slight left ventricular enlargement. There was a small left-sided cervical rib. X-ray of the hands showed moderate shortening of both fifth metacarpals. An ECG showed an rSr' pattern in V1 only. Catheterization showed a gradient of 25 mmHg across a coarctation at the usual site, which was also demonstrated angiographically; and despite the mildness of the gradient the latter investigation showed well-marked post-stenotic dilatation of the aorta distally. Seven years later she had an uneventful first pregnancy and confinement.

Discussion

Van der Werff ten Bosch (1959) indicated that the condition known as pseudo-pseudo-hypoparathyroidism had never previously been adequately defined, and proposed that the term be reserved for patients with short stature and shortened metacarpals, without tetany. He suggested that the term brachymetacarpal dwarfism (BMD) be used as an alternative and more descriptive designation; and certainly this is not only much less cumbersome, but also a much less misleadingly remote derivation from the original stem-entity of hypoparathyroidism

(Gibson, 1961). His report of eight cases of this condition included four with features of Turner's syndrome as well, and his seventh case had hypertension, but no clinical details as to its cause were given. An exact definition of what constitutes shortness of stature, or even dwarfism, is difficult to find in a search of the literature (although some textbooks of endocrinology naturally refer to dwarfism in detail), but as far as female height is concerned probably 158 cm (5 feet 3 in) is acceptable as the lower limit of normal, and has been so regarded in the seventh case described by van der Werff ten Bosch and the second case described above. Many authors have commented on an association between Turner's syndrome and BMD (Nagant de Deuxchaînes, 1959; Mann, Alterman & Hills, 1962; Schwarz, 1964; Gardner, 1970). It seems reasonable to suppose that there is a clinical spectrum of these two conditions, ranging from pure Turner's syndrome to pure BMD, with overlapping examples between the extremes. Coarctation is a well-known complication of Turner's syndrome, occurring in from 20 to 35% of cases according to different authorities. Its association with BMD does not yet seem to have been described, but its occurrence may have been underestimated in cases of pure BMD lacking any features of an associated Turner's syndrome which might draw attention to the cardiovascular system—any mild coarctation being easily overlooked. In discussing the aetiology of coarctation Campbell & Polani (1961) analysed 151 cases from various points of view—including non-cardiac malformations in the propositi and their relatives—but in none of them were any features mentioned suggesting BMD. Clinical screening for the latter condition in coarctation is easily done by looking for dimpling of any knuckles on clenching the fists (Albright's 'first sign') or general smallness of either the hands or feet or both. If the patient is also a dwarf then the diagnosis is confirmed according to the criteria proposed above; although additional features such as facial rounding, mental subnormality, subcutaneous calcification, or exostoses can occur as well.

The above cases are reported in order to encourage further clinical search for such association between BMD and coarctation, and thereby possibly obtain enough evidence to warrant their definition as a clinical syndrome.

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- Clinical Aspects of Dementia*. By J. PEARCE and E. MILLER. Pp. 142, illustrated. London: Bailliere Tindall and Cassell. 1973. £3.00.

Book reviews

Abdominal and Vaginal Hysterectomy—New Techniques based on Time and Motion Studies.

By S. JOSE COHEN. Pp. 170, illustrated. London: William Heinemann Medical Books, 1972. £3.50

The author of this outstanding book has managed to solve two most difficult problems. In the first place he has written a wonderfully interesting technical account of two operations, each described in a manner both simple and lucid. In the second place he has succeeded in conveying his own convictions of the intrinsic value of the special procedures he has perfected in the course of his own long experience.

For these two reasons, *Abdominal and Vaginal Hysterectomy* must be regarded as an essential part of every gynaecologist's library. Wholly readable, it will provide food for thought for all save those who steadfastly refuse even to contemplate any alterations or improvements in their own operative technique and dexterity. It is, in fact, one of the most valuable publications of recent years in this particular specialty since it contains information which cannot be found elsewhere.

Neuropharmacology and Behaviour

By V. G. LONGO. Pp. 184, illustrated. Reading, England: W. H. Freeman, 1972. £3.30 (paperback £1.70).

The gulf between the pharmacologist and clinical psychiatrist cries out to be bridged. A number of authors have attempted this feat with little success, and this volume is no exception, leaving the reader poised in mid-stream. By a pharmacologist from the Institute Superiore di Sanita, Rome, it is attractively printed and set out in enticing sections on Anti-Psychotics, Anti-Depressants, Tranquillizers and Hallucinogens. Longo gives a concise account of the pharmacology and the clinical application of neuroleptic drugs and a more detailed account of some obscure hallucinogens which might be of interest to undergraduates, and those requiring a review of the subject. Although the book is commended by the publishers for 'medical, psychiatric, and pharmacological researchers', and as a reference work, it has no comprehensive index (though there are useful references at the end of each section). A 'glossary-index' is appended in which some commonplace medical terms as clonic, carotid, hyperpyrexia, dysuria, myocardial and hallucination are defined.