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EDITORIAL

BONE REMODELLING

A bone is a wonderful thing. A long bone starts life as a bar of mesenchyme, without strength or form. It develops eventually into a light, tubular structure of immense strength, narrower in the middle than at the ends, and specially designed to withstand the enormous stresses and strains of normal daily activity. The centre becomes hollowed out to allow lightness and to act as a storehouse for marrow. Basic structure consists of strands of protein matrix impregnated with apatite (calclum triple phosphate with some carbonate), rather like reinforced concrete, which are laid down according to precise mechanical laws governed by the needs for strength in particular directions to withstand the prevailing forces.

As originally laid down from rows of cartilage cells the new bone consists of rather dense longitudinal strands of calcified osteoid--the primary trabeculae. The osteoclasts now play a major part in the remodelling of these trabeculae. (Let us leave aside the question of whether osteoclasts really directly 'eat away' bone—it is easier to consider them as doing so.) While osteoclasts remove portions of these trabeculae, osteoblasts will lay down more bone as needed. A particularly pretty example is the growth of a rib: osteoclasts remove bone from the inner sides of trabeculae while more bone is laid down on the other sides. In the long bones the external effect of this remodelling is the absorption of outer layers of bone, so as to produce the tapering which occurs as we travel from the epiphysis along the shaft. If this were not so an adult femur, for instance, would be the same width in the secondary trabeculae of the cortex, the spongy cancellous bone of the medulla and the actual marrow cavity.

In this intricate process of bone development it is natural that things may go wrong, and lead to disorders of remodelling. In these states the lower end (or metaphysis) of the femur shows well the abnormality in shape, which has been likened to an Erlenmeyer flask. The best known clinical condition is that of osteopetrosis (Albers-Schönberg, or marble bone disease). The bones are over-dense, probably because of deficient activity of osteoclasts. With high-penetration X-rays longitudinal lines may be seen which represent the non-remodelled primary trabeculae. Further

VAN DIE REDAKSIE

BEENVERVORMING

Been is iets wonderliks. In die begin bestaan 'n langbeen uit vrugbindweefsel en is dit sonder vorm of krag totdat dit uiteindelik in 'n ligte, pypvormige struktuur ontwikkel wat geweldig sterk is; dit is dan wyer aan die ente as in die middel en is spesiaal bestem om die geweldige eise van die normale daaglikse lewe te weerstaan. Die binneste is hol—derhalwe is die struktuur lig en is daar ook plek vir die murg. Basies bestaan die struktuur uit proteïenmatrysdrade wat met *apatite* (kalsium drievoudige fosfaat en 'n bietjie karbonaat) deurtrek is, wat, ietwat soos gewapende beton, ooreenkomstig streng meganiese wette gelê word al na gelang die noodsaaklikheid om in spesiale rigtings weerstand te bied.

Die nuwe been wat oorspronklik uit rye kraakbeenselle opgebou is, bestaan nou uit taamlik digte longitudinale verkalkte *osteoid*-drade—die primêre trabeculae. Beenvretende selle speel nou 'n groot rol in die vervorming van die trabeculae. (Ons behandel hier nie die vraagstuk of beenafbreekselle in werklikheid die been direk ‚wegvreet' of nie—dit sal makliker wees om aan te neem dat dit wel die geval is.) Terwyl dele van hierdie trabeculae deur beenafbreekselle verwyder word, word meer been deur beenvormende selle gebou al na gelang die vereistes. Die groei van 'n rib is 'n besonder mooi voorbeeld hiervan—been aan die binnekant van die trabeculae word deur beenafbreekselle verwyder terwyl meer been aan die buitekant gelê word. Die uitwendige uitwerking van hierdie vervorming van die langbene is die absorpsie van die buitenste beenlae sodat die been geleidelik dunner word van die epifise langs die pyp af—as dít nie die geval was nie, sou die pyp en die onder-ent van 'n volwasse dybeen byvoorbeeld ewe breed wees. Inwendige vervorming is verantwoordelik vir die digte sekondêre trabeculae van die skors, die sponsagtige been van die medulla en die werklike murgholte.

Met so 'n ingewikkelde proses van beenvorming is dit te verwagte dat dinge verkeerd kan gaan en steurings kan voorkom. Onder sulke omstandighede toon die onderste gedeelte (of metafise) van die dybeen die abnormaliteit in vorm duidelik—dit kan met die vorm van 'n Erlenmeyer-fles vergelyk word. Die bekendste kliniese toestand is dié van osteopetrose (Albers-Schönberg of marmerbeensiekte). Die been is te dig, waarskynlik omdat die osteoklaste nie bedrywig genoeg is nie. Met dieppenetrerende X-strale kan lengtelyne gesien word wat die nie-vervormde trabeculae verteenwoordig. Verdere interessante radiologiese bewyse van

interesting radiological evidence of lack of remodelling occurs in the frequent appearance of the outlines of smaller bones within bones; for instance a foetal-sized phalangeal shadow within the present phalanx. Further, the marrow cavities are exceedingly small, and the bones, despite their density, are liable to fracture because their substance has not been laid down in a sound mechanical manner.

Less severe lack of modelling is seen in the condition described by Pyle [1] as 'familial metaphyseal dysplasia', in which the metaphyseal regions are wide, as in osteopetrosis, but there is no excessive bone density, the cortex is thin, and the narrow cavity is reduced in size. Fractures may occur, and the condition appears to be inherited as a Mendelian dominant characteristic. In one family idiopathic osteoporosis accompanied it.[2] In a few cases this metaphyseal dysplasia has been combined with a severe and disfiguring overgrowth of bones of the skull and face—a variety of leontiasis ossea. This combination has been called 'cranio-metaphyseal dysplasia'.[2]

Other states which are associated with deficient bone remodelling and flask-like lower femora include the infiltration of Gaucher's disease and the marrow hyperplasia of the congential haemolytic anaemias, especially Cooley's type. Vitamin-A deficiency also produces a lack of remodelling in experimental animals, but plays no known part in the human syndromes.

Defects in remodelling with increased width in the mataphyseal regions can also be seen in other developmental osseous dystrophies such as multiple exostoses (diaphyseal aclasis of Keith) or enchondromatosis (Ollier's disease). In these, as in some other congenital dysplasias, the metaphyseal abnormality is plainly not of primary importance, and is here produced by a disruption in development due to the displaced islets of cartilage cells which appear in these regions. Finally temporary deficiency of remodelling may occur in various states such as lead poisoning and scurvy.

1. Pyle, E. (1931): J. Bone Jt. Surg., **29,** 874.
2. Jackson, W. P. U., Albright, F., Drewry, G., Hanelin, J. and Rubin, M. I. (1954): Arch. Intern. Med., **94,** 871.

'n vervormingsgebrek kan herhaaldelik gesien word in die voorkoms van buitelyne van kleinere bene binne in die bene; byvoorbeeld die skaduwee van 'n fetaalgrootte toonbeentjie binne die werklike toonbeentjie. Verder is die murgholtes besonder klein, en die bene ten spyte van hul digtheid, is geneig om te breek omdat hul stof nie op 'n deeglik meganiese manier gelê is nie.

'n Minder ernstige vervormingsgebrek word gesien in die toestand wat Pyle[1] as ‚familial metaphyseal dysplasia' beskryf—soos in osteopetrose is die metafisestreke uitgebrei, maar daar is geen uitermatige digtheid van die been nie, die skors is dun en die nou holte verklein. Breuke kan voorkom en dit blyk asof die toestand as 'n Mendeliaan-dominante eienskap oorgeërf word. In een familie het dit met idiopatiese osteoporose gepaard gegaan.[2] In 'n paar gevalle is hierdie ‚metaphyseal dysplasia' vergesel deur 'n ernstige en skendende beenaangroeisel aan die skedel en gesig ('n varieteit van leontiasis ossea) waarvoor die benaming ‚craniometaphyseal dysplasia' al gebruik is.

Ander toestande wat met beenvervormingsgebrek en flesvormige onderste dybene geassosieer is, is die infiltrasie van Gaucher se siekte en die murghiperplasie van aangebore hemolitiese bloedarmoede, veral die Cooleytipe. 'n Tekort aan vitamien-A lei ook by proefdiere tot vervormingsgebrek, maar sover ons weet word dit nie in die menslike simptome-groep gevind nie.

Vervormingsgebreke met verdikking van die metafisestreke, kan ook gesien word in ander distrofië van die ontwikkelende been soos in veelvoudige eksostoses (*diaphyseal aclasis* van Keith) of enchondromatose (Ollier se siekte). In hierdie, soos ook in sommige ander aangebore dysplasië, is die metafise-abnormaliteit definitief nie van die grootste belang nie en word dit veroorsaak deur 'n onderbreking in die ontwikkeling wat te wyte is aan verplaasde kraakbeensel-eilandjies wat op hierdie plekke voorkom. Ten slotte kan 'n tydelike vervormingsgebrek by toestande soos loodvergiftiging en skeurbuik voorkom.

1. Pyle, E. (1931): J. Bone Jt. Surg., **29,** 874.
2. Jackson, W. P. U., Albright, F., Drewry, G., Hanelin, J. en Rubin, M. I. (1954): Arch. Int. Med., **94,** 871.

BEDSIDE DIAGNOSIS IN HEART DISEASE

Heart disease has been generally regarded as a branch of medicine in which the arts of bedside diagnosis could be fully exercised. Sir James McKenzie showed how clinical study of the arterial and venous pulses, palpation of the praecordium and diligent auscultation would lead in most cases to an adequate diagnosis. Later the acumen of the physician was supplemented when Sir Thomas Lewis brought electrocardiography to the clinic and Sir John Parkinson established cardioscopy as an essential part of cardiac examination.

In 1939, when cardiac surgery became a reality and the demand for precise pre-operative diagnosis arose, complicated mechanical techniques were elaborated and for a time it seemed that the clinical arts would be washed away by a tidal wave of cardiac catheters, angiocardiograms, phonocardiograms, vectorcardiograms and ballistocardiograms. Fortunately this was a temporary phase and from the technical jungle a new appreciation of bedside investigation in heart disease has emerged. New physical signs have been discovered, many old ones have been evaluated again and invested with a new significance, and accurate *clinical* assessment of the most complicated structural and functional disorders of the heart has become possible.

In this connection we are pleased to note the significant contribution to bedside diagnosis recently published by Dr. V. Schrire and Dr. L. Vogelpoel from the cardiac clinic at Groote Schuur Hospital, Cape Town.[1] They have investigated many cases of tachycardia with regular rhythm and have shown how supraventricular tachycardia may be readily distinguished clinically from ventricular tachycardia.

In supraventricular tachycardia, as in the normal heart, there is only slightly asynchronous ventricular

contraction; the heart sounds are therefore minimally split. In ventricular tachycardia, where there is an ectopic pace-maker in one ventricle, ventricular contraction is markedly asynchronous and the splitting of the heart sounds becomes abnormally wide.

The intensity of the first heart sound depends on the position of the cusps of the auriculo-ventricular valve at the beginning of ventricular systole. In ventricular tachycardia there is dissociated auricular and ventricular activity and so the position of these cusps varies from cycle to cycle and the first heart sound accordingly has a varying intensity. As a further consequence of this dissociation, the right auricle often contracts against a closed tricuspid valve, producing large, irregular, independent 'cannon waves' in the jugular venous pulse. In supraventricular tachycardia, on the other hand, there is no auriculo-ventricular dissociation; the first heart sound therefore is of constant intensity and irregular venous 'cannon waves' do not occur.

Attention is drawn to the difficulty in distinguishing on a routine electrocardiogram between supraventricular tachycardia with bundle-branch block and ventricular tachycardia. The distinction is important because vagal stimulation or digitalis may be necessary for the former, whereas quinidine or procaine amide is the treatment of choice in the latter. The physical signs, however, may be more helpful than the electrocardiograph. In both cases, there is abnormally wide splitting of the heart sounds but in the supraventricular rhythm there are no irregular venous 'cannon waves' and the first heart sound is of constant intensity.

The authors are to be congratulated on the first publication from this newly-established clinic; further contributions will be anticipated with much interest.

1. Schrire, V. and Vogelpoel, L. (1955): Amer. Heart J., **49**, 162.