

PAEDIATRIC VIEWPOINT

What exactly is paediatrics? Why is it spelled that way? Is it a separate 'discipline' to be taken seriously or just the result of a pressure-group serving its own ends? Is the slightly derogatory term of reference 'kids', used by students and nurses, a reflection of widely-held opinion? Or is paediatrics something worse, to be suppressed as much as possible, since it threatens to claim a place in the front rank of medicine and endanger the already diminishing status and shakily-entrenched privilege of hierarchies of physicians, surgeons, and obstetricians? If it is important, what are the medical schools, nurses' training centres, and public health authorities doing about it? And what do paediatricians discuss at their congresses — exercises in the occult or elaborate techniques for the changing of diapers? Why produce a paediatric number of the *Journal*?

These questions are provoked by the attitudes of those people, lay and medical, with whom paediatricians are in daily contact. There is a feeling of suspicion about the whole business, of paediatrics becoming an *enfant terrible* in this country. It clamours for more teaching time, more beds, more nursing and auxiliary services, for special hospital facilities, for convalescent homes, and of course for more money for its activities and research. And it is not the only one of medicine's 'problem children'. There are others growing up and, sooner or later, they also will have to be admitted to the limelight from their present semi-obscurity in the wings. The processes of evolution are not confined to zoology, and paediatrics in South Africa has a long way to go. In many other respects, notably virology, nutrition, and cardiac surgery, this country's experts are of international status and are accorded approval at home. Much of their work has been done in the treatment of children, but the basic requirements of paediatrics continue to be largely ignored.

Dorland's *Illustrated Medical Dictionary* defines paediatrics (or rather, American fashion, 'pediatrics') as 'that branch of medicine which treats of the child and its development and care and of the diseases of children and their treatment'. The word is derived from the Greek *pais*, *paidos*, a child, and the American way of spelling it has probably led to the confusion in some people's minds that paediatrics deals with diseases of the foot (from the Latin, *pes*, *pedis*, a foot). But the confusion does not end there, since the general vague mental picture of a paediatrician is that he is merely a 'baby doctor' and the inference is that he should be treated as such, the baby being parsed as an adjective and not as a noun. The care of children is the common lot of a very large proportion of the human race at some time in their lives, but their equipment for the job is negligible. This applies to medical and nursing personnel almost as much as to the layman. There is vast and urgent need for expert paediatric knowledge and guidance in every type of medical institution, except those for alcoholics, and there is no dis-

cernible recognition of this at present. The lay public is still in paediatric Standard I.

Approximately a third of any general practice is paediatric work in terms of the definition above and it is not many years since the statement was made that 'I'd as soon see the devil himself come into my consulting room as a young child'. The era of castor oil as a panacea for all ills in childhood has not yet passed and the prophylaxis of disease in childhood is an unopened book for large sections of our population of all races. The special requirements, meticulous attention to hygiene, detailed care and observation, psychological needs of sick children, and what the Americans call T.L.C. (tender loving care) in every aspect of the work, are fundamental. But they have yet to make the faintest impression, let alone any sort of impact, on the vast majority of those who hold the children's future in their hands. And, though it is almost incredible, this applies to women even more than to men. In other civilized countries paediatrics has been an established branch of medicine for at least forty years and much of the earliest work leading to modern medical practices was first done in the treatment of children by paediatricians or their immediate associates. Fluid replacement and biochemical investigations were widely used in paediatric circles long before they were applied to adults, and the work of Hartmann, Gamble and Darrow, and others was done in children. But in South Africa, in 1960, there are only two professors concerned with paediatrics, a few more lecturers in charge of university departments, a mere handful of paediatric-trained nurses and no sign of any disposition to increase this miserably token force.

Far too many of our children, in and out of the hospitals, are cared for according to the tenets of their own or someone else's grandmother. Medical students and postgraduates, these days, have a modicum of instruction in paediatrics. As interns, they will be helped by nursing staff, if they have trained nurses to care for their patients, who will largely follow the diets and practices of their mothers, who were instructed in the art by *their* mothers, half of whose children died before the age of five years. The less fortunate doctors, and their paediatric patients, will be at the mercy of individuals trained, or in the process of being trained, by instructresses who are themselves untrained in paediatric nursing. Or, and this is not unknown, the attendants will be untrained and often untrainable. Folk-lore has been tried over centuries, with deplorable results. Surgeons do not have to work with untrained staffs in pre-Listerian conditions. Physicians treating adults achieve their effects by drugs and the cooperation of the patient. Obstetricians, to some degree, are at the mercy of their staff and surroundings for the safety of their adult patients, and in exactly the same position as paediatricians otherwise. In other words, the needs of one branch of the profession are much the same as those of the others. But when the patients cannot, because of age or illness

or merely from timidity, voice their needs and complaints, the deficiencies in our ritual become glaringly obvious. In paediatrics they became so long ago, and it was realized that the maintenance and the restoration of health in children, still more in infants and in the newborn, demanded far more particular knowledge and attention to detail than was needed in other fields of medicine. This applies to dietetics, hygiene, environment, and even to clothing.

Diseases of children differ in some degree from those of their elders partly because of the size factor and partly because of the greater incidence of non-degenerative types of illness. The outlook of the paediatrician is towards either the maintenance or the complete restoration of health, as opposed to the preservation of what health remains in most adult patients, or the repair of some

form of damage as in most of surgery. To that extent paediatrics is a different but not an alien discipline. It recognizes the need to cooperate with many other divisions of medicine to achieve its aims but it is not a 'very small subject' capable of being absorbed by any other. The highest standard of paediatrics is possibly Utopian. The current general standard, in all kinds of ways, could and must be improved. Until it is, it is to be hoped that paediatricians will continue to be agitators.

The present number of the *Journal* should serve to show that the professional interests of paediatricians are in line with what has been indicated above. The discussions at the recent paediatric congress have a bearing on many medical problems, though here their background is the paediatric age group.

### RENALE HIPOFOSFATEMIE

Sedert die publikasie van Richard Bright se referaat in 1827 was dit gebruiklik om die term chroniese renale ontoereikendheid te assosieer met die kliniese en patologiese bevindings soos deur hom beskryf. Die patogenese van die stikstofretensie, albuminurie, metaboliese asidose, en anemie is nog nie in elke opsig verduidelik nie, maar daar is algemene ooreenstemming dat ons hier te doen het met versaking van die nierfunksie in sy geheel. Daar is 'n vermindering van die aantal funksionerende nefrone met 'n ooreenstemmende daling van die filtrasie deur die glomeruli asook verstoring van die tubulêre funksies, soos gevind by gevalle van chroniese glomerulonefritis, arteriële nefrosklerose, en chroniese piëloefritis.

GEDURENDE die laaste paar dekades het 'n toenemende aantal navorsers intensiewe aandag gewy aan 'n groot verskeidenheid van renale verstorings wat in teenstelling staan met die sg. Bright se siekte.<sup>1,2,3</sup> Hierdie toestande is in die eerste instansie gekenmerk deur tekens van renale ontoereikendheid veroorsaak deur defekte van die tubulêre funksies. Albuminurie en stikstofretensie is gewoonlik afwesig, hoewel glomerulêre ontoereikendheid uiteindelik mag ontwikkel. Die teenswoordige belangstelling in die laasgenoemde groep van siektetoestande kan toegeskryf word aan verskeie faktore, byvoorbeeld die feit dat die toestande behandelbaar en die skade gewoonlik omkeerbaar is, in teenstelling met chroniese nefritis. Die sindrome bied ook 'n unieke geleentheid aan om renale meganismes en geneties-bepaalde siektetoestande te bestudeer. Baie van die sindrome gaan gepaard met aminosurie, en die onlangse ontwikkeling van chromatografiese en ander tegnieke om aminosure wat in die urine afgeskei word te bepaal,<sup>4</sup> het as 'n verdere stimulus gedien om belangstelling te bevorder.

Die patogenese van die siektetoestande wat veroorsaak word deur gebrekkige funksie van die renale konkelbuïes berus op defektiewe herabsorpsie van stowwe in die glomerulêre filtraat. Byvoorbeeld, defektiewe herabsorpsie van (a) water gee aanleiding tot nefrogeniese diabetes insipidus,<sup>5</sup> (b) glukose → renale glukosurie, (c) fosfate → renale hipofosfatemie (d) glukose, fosfate, en aminosure → Fanconi se sindroom, en (e) alkalië → renale asidose. So is daar nog etlike ander defekte en aangesien amper enige *kombinasie* van defekte oënskynlik kan plaasvind,

soel as *enkele* defekte, kry ons 'n groot aantal verskillende interessante siektetoestande, die meeste waarvan selde voorkom. Renale hipofosfatemie is een van die algemeenste.

Die terminologie van hierdie siektes is menigvuldig en taamlik verwarrend. Fanconi se naam word soms losweg gebruik om 'n hele paar sindrome in die groep te bestempel. Dit behoort beperk te word tot (d) hierbo. De Toni en Debré se name behoort na regte bygevoeg te word aangesien hulle ook oorspronklik dergelike gevalle gerapporteer het. Eintlik sou dit verkieslik en minder verwarrend wees om name te vermy soos verskillende mense, o.a. Fanconi self, alreeds aangedui het.

Wat renale hipofosfatemie betref, word 'n dosyn of meer terme gevind in die literatuur om dieselfde siektetoestand aan te dui. Die benaming vitamien D-weerstandige rachitis<sup>6,7</sup> is die populêrste. Daar is egter ander tipes van rachitis wat vitamien D weerstandig is. Die toestand is nie 'weerstandig' as massiewe dosisse van die vitamien toegedien word nie, en daar is geen weerstandigheid teen die vergiftigingseffekte nie. Die term is dus nie baie aanneemlik nie.

Al die bevindings by hierdie gevalle kan verduidelik word op grond van defektiewe funksie van die proksimale renale konkelbuïes. Oënskynlik is daar 'n defek van die ensiemsisteme wat die transport van fosfate beheer. Die tubulêre defek is gewoonlik, indien nie altyd nie, kongenitaal. Die swaanek-deformiteit van die proksimale tubulêre stelsel<sup>8</sup> wat gevind is by verskeie gevalle van verwante renale verstorings, is oënskynlik nie die patologiese letsel wat in verband staan met renale hipofosfatemie nie.

Ander faktore wat 'n rol speel by die patogenese van hierdie siekte moet oorweeg word. Albright<sup>9</sup> het gemeen dat hiperparatiroidisme 'n hoofrol speel en het dan ook hiperplasie van die paratiroidklier aangetoon. Hierdie hipotese verklaar egter nie al die feite nie. Die effek van vitamien D op die funksionele bevoegdheid van die proksimale konkelbuïes het die laaste tyd baie aandag geniet. Fanconi stel renale hipofosfatemie of weerstandige rachitis aan die een uiterste van 'n spektrum van siektetoestande wat gekenmerk word deur verskillende grade van gevoeligheid vir vitamien D. By renale hipofosfatemie sal daar

dan die laagste graad van gevoeligheid wees teenoor vitamien D. Chroniese hiperkalsemie van suigeling kom aan die ander end van die spektrum, en word beskou as 'n toestand wat veroorsaak word deur oorgevoeligheid vir hierdie vitamien.

Die skelet self se aandeel in die patogene is oënskynlik heeltal passief. Verwyderde beenweefsels van pasiënte sal kalsifiseer in die teenwoordigheid van kalsium en fosfor. Die bene kalsifeer nie as hul geïnkubeer word met die ongewysigde serum van onbehandelde pasiënte nie.

Die siekte is oorerflik op 'n dominante seksverbonde genetiese basis. Winters en sy medewerkers,<sup>10</sup> byvoorbeeld, het 'n familie bestudeer na hul die siekte by 'n kind ontdek het. Daar was 283 lewende familieledes, waarvan driekwart ondersoek is. Daar was rachitiese deformiteite teenwoordig by 25 en hipofosfatemie by 11 sonder deformiteite. Kliniese ondersoeke sonder meer is dus nie voldoende nie. Hulle het gevind dat daar geen geval van die siekte voorgekom het waar die ouers alteele normofosfatemies was nie. Egte sporadiese gevalle word egter af en toe gerapporteer.<sup>11</sup>

Al die kliniese verskynsels kom voor in die skelet. Deformiteite word gewoonlik bespeurbaar as die kind begin te loop. Die kenmerkende tekens van rachitis is teenwoordig en röntgenografiese ondersoeke toon ook die tipiese rachitiese verskynsels. Die pasiënt verkeer andersins in blakende gesondheid en het 'n normale lewensverwachting, in teenstelling met dié tipe van renale rachitis wat veroorsaak word deur chroniese nierversaking. Op grond van bogenoemde bevindings word die kind, redelik genoeg, behandel as 'n geval van rachitis met die gewone terapeu-

tiese dosis van vitamien D. Ongelukkig het daar in die verlede gewoonlik 'n tydperk van gemiddeld twee jaar verloop voor die geneesheer bewus geword het van die weerstandige aard van die siekte, en gedurende hierdie tyd ontwikkel uitgesproke deformiteite en dwergisme.

Behandeling met massiewe dosisse van vitamien D, soms soveel as een miljoen eenhede per dag, of 25 mg. calciferol, is van beproefde waarde, maar die geneesheer moet onthou dat sulke dosisse giftig mag wees en die aanbevole voorsorg moet noukeurig nagekom word.<sup>2</sup> Die behandeling gaan 'n onbepaalde aantal jare lank voort. Na puberteit sal dit waarskynlik moontlik wees om die dosis te verminder.

Baie gevalle is in die verlede oor die hoof gesien en medici moet op hul hoede wees om die siekte vroeër te diagnoseer en om te onderskei tussen gewone rachitis weens eksogene oorsake, renale rachitis wat voorkom by gevalle van chroniese nefritis met hiperfosfatemie, en die tipes van renale rachitis wat veroorsaak word deur verstoring van die tubulêre funksies.<sup>2</sup>

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## PAEDIATRIC SURGERY

That well-known story in which the elderly practitioner inquires of the fledgling otorhinolaryngologist which nostril he is going to specialize in, emphasizes the often ridiculously high value placed nowadays on specialization. But in this latter half of the twentieth century, with rapid advances being made almost daily in all branches of science, including medicine, it is becoming increasingly apparent that the specialist is here to stay. Almost every organ in the adult now has its expert, whereas the child, especially the surgically-ill child, for too many years has had deaf ears turned to its insistent cry. It is interesting indeed that paediatric surgery, which only in the last few decades joined the family of specialties, has now grown into a lusty 'infant', howling for recognition and demanding a place at the table.

There are many reasons for the earlier cruel neglect of paediatric surgery; e.g., the patient is inarticulate, there is a widespread misconception that children can be treated simply as miniature adults, and the fact that the salvage rate of congenitally-deformed infants has been low. In addition the financial position of the little patient is often precarious because his young parents are on the threshold of their married partnership and usually in no position to afford the fees of a specialist.

There is little to compare with the satisfaction obtained in remedying an otherwise fatal neonatal anomaly and then seeing the infant thrive and watching, with the happy parents, the child growing up. Where in surgery can one expect a sixty to seventy year prognosis, other than in the surgery of infancy?

A demand has been created for experienced surgeons who will be prepared to devote all their time to paediatric surgery. This demand is now being met. All over the world more and more men are training to become proficient in this branch of surgery—an exacting branch, requiring specialized knowledge of the anatomical, physiological, metabolic, and psychological needs of children, and demanding the utmost gentleness, skill, and precision in operative work. The knowledge and the skill can only be acquired by constant daily contact with small children, close personal pre- and postoperative supervision, and frequent experience in operating on small infants. Training of this nature is long, and the aspirant paediatric surgeon must have a great love of children, from which will surely stem a deep desire to obtain and give his best for the child who is in need of surgical care.

The training of specialists in paediatric surgery can only be accomplished in a children's hospital, where, under the guidance of a senior surgeon (whose primary, if not total, interest is children's surgery) the proper practice of the subject can be taught. In such a hospital, which must be fully staffed and adequately equipped, the apprenticeship will be a long one. It should follow a period of training in general surgery, including the major subdivisions of orthopaedics, plastic surgery, urology, and thoracic and cardiac surgery. For the paediatric surgeon is a modern example of that almost archaic species, the general surgeon. His interests range from clubfeet to naevi; from undescended testes to holes in the heart; from oesophageal atresia to enuresis.

In South Africa, too, paediatric surgery is growing up, thanks largely to the interest accorded it during the last ten years in Cape Town. At the Red Cross War Memorial Children's Hospital in Cape Town the high standard of paediatric surgery compares favourably with that of similar centres in the United Kingdom, the United States, Switzerland and Denmark—indeed anywhere in the world. There is a full-time consultant surgeon, unique in South Africa, whose time is devoted entirely to paediatric surgery, and there are registrars who maintain a night and day watch on all major neonatal surgical cases. No mention of paediatric surgery in South Africa would be complete without coupling it with the name of Prof. J. H. Louw of the University of Cape Town, who has always strenuously exerted himself in the cause of this demanding

specialty.\*

It is only by whole-hearted and full-time attention that unnecessary loss of life can be prevented, and the best results obtained. The day of the casual paediatric surgeon, operating on infants, perhaps once a year, knowing no more about their special care than about the metabolism of magnesium, is fortunately fading. With the widespread, ever-increasing awareness of the importance of paediatric surgery, the consequences cannot but be beneficial to thousands of tiny patients to whom our duty is crystal-clear—to preserve that most precious thing in the world, life itself.

\* A paper on 'Surgery in the newborn' by Prof. J. H. Louw, which was presented at the Fourth Congress of the South African Paediatric Association (M.A.S.A.), Cape Town, 5 April 1960, was published in the issues of the *Journal* for 13 and 20 August 1960 (34, 686 and 707).