The Bulletin of the
Kandang Kerbau Hospital,
Singapore.
Vol. IV—No. 2—October 1965

Neonatal Orthopaedics

by

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Lecture delivered in October 1963 to Obstetrical & Gynaecological Society, Singapore.

In the newborn, the orthopaedic problems fall into three main groups: birth injuries, cerebral palsy and congenital skeletal abnormalities, all of which may give rise to a variable degree of dysfunction.

Birth fractures and other injuries at delivery are inevitable in certain circumstances but their management is straighforward and these babies are quickly transferred to orthopaedic care.

Children with cerebral palsy do not often present to the orthopaedic surgeon until later in infancy; they then provide many difficulties indeed for their mechanical problems have to be overcome often in the face of insufficient intellectual function as well as abnormal motor control. The management of the locomotor defects in cerebral palsy is concerned with two objects; to obtain the best possible functional use of the limbs and to prevent or correct deformity. Later, in the treatment of potential or established deformity, a wide range of orthopaedic procedures are available.

Congenital skeletal deformities are of two kinds; primary, due to errors of the germ plasm and its development; and secondary, due to stresses acting on skeletal tissues that have developed normally. These stresses may be mechanical or due to abnormal biochemical or hormonal influences on the foetal metabolic environment.

Skeletal disorders are difficult to classify; in many instances their cause is obscure, knowledge of the pathology limited, and long-term information on the natural history of many conditions is lacking. Furthermore, often the skeleton is simply a participant in a disordered body as a whole.

This simple classification will help to indicate those more common disorders which the orthopaedic surgeon may find of interest.

1. The large group of dysplasias and bone dystrophies

Strictly speaking, dysplasia means faulty development or growth and dystrophy implies faulty nutrition. Tradition however, allows a wider use of these terms. The disorders of growth include achondroplasia, osteogenesis imperfecta, diaphyseal aclasia, osteo-chondro dystrophy etc. The second half of the group do not really have any significance in infancy, they are the disorders of bone density. Bone density is deduced from radiographs and is a measure of the amount and distribution of the mineral content of bone. These conditions include osteopetrosis and osteopoikily. Although it is something of an oddity, arthrogryposis may be inserted here for it is dependant on the faulty differentiation of muscle giving rise to extensive disability. The dystrophies and dysplasias are almost entirely due to disorders of the germ plasm being often present in more than one member of a family.

2. Errors of segmentation

Fused segments may be represented by fusion of vertebral bodies more commonly seen in the upper dorsal and cervical spines. The most common manifest syndrome is that of Klippel-Fiel. There may be additional segments for example, supplementary digits, and suppressed segments are seen as ectromelia and ectrodactaly. Single muscles may be absent, most commonly the pectoral muscles but this may be due to the obvious deficiency which results. Segmentation may take place at an

abnormal level or there may be a failure of a segment o assumet its proper level, as in Sprengel's shoulder.

3. Errors of ossification

These must not be confused with disorders of bone density. In this group we include spina bifida and myelomeningocele which may provide immediate or delayed problems for the neurosurgeon as well as his orthopaedic colleague. Extensive spina bifida is often associated with complex orthopaedic conditions of the lower legs particularly the feet. The prognosis for these children has constantly improved. Provided progressive hydrocephalus does not occur, the majority of these children have a normal or near normal intelligence. In addition they have normal upper limbs. Consequently a large number of these children have the potentiality of walking and therefore require orthopaedic management from the time of their birth. The abscence of complete bones for example, the radius, fibula or femur gives rise to dreadful disability which may lead to amputation in early life.

4. Congenital dislocation of the hip

This is of such importance in many parts of the World that it merits separation. Emphasis has been placed here on the importance of early diagnosis and treatment and in no condition is this more pertinent than in congenital hip dislocation. In Manchester, Barlow (1962) described his investigation of over nine thousand babies in whom he found and treated one hundred and nineteen abnormal hips at birth. All but twelve of these were stable within two months. The remaining twelve were all clinically normal with concentric hips at one year and were walking normally. These figures serve as an illustration of the results which are possible if the new-born are routinely checked, for they provide an impressive contrast with the group of one to three years olds with classical congenital dislocation of the hip with which we are all too familiar in Europe. These results are of course, a reflection of the techings of von Rosen in Sweden. In Malmo where he practises, all the babies are delivered in hospital where the midwives are taught to look for dislocation of the hip at birth so that treatment is instituted at once. As a result of these methods he has been able to claim that no new case of established dislocation has occurred in Malmo during the period of the investigation.

5. Talipes or club-foot

Many of the remarks applied to congenital dislocation of the hip are equally applicable in the treatment of talipes. Manipulative correction in the first twenty-four hours of life and simple maintainence of this position reduces the problem of talipes to those few 'malignant' feet which seem not to respond to simple measures. Those cases treated early have every prospect of normal posture and gait at a normal time. Again the comparision with the unfortunate children who are not brought for treatment until several months old is adequate testimony in favour of early attention. Talipes is an orthopaedic emergency in the newborn.

6. The metabolic disorders

These include diabetes and vitamin-D resistant rickets but unless one has previous knowlege of the possibility of transmitted abnormality the diseases in this group are not usually appreciated in the neonate.

7. Disorders of organisation

These are mainfested by certain forms of syndactaly, microdactaly and intra-uterine amputations.

Unless babies are routinely examined by a paediatrician in the first day of life—which is the ideal arrangement—it is important for the obstetrician to recognise those instances where early orthopaedic assistance can be of value. If skilled help is not readily available then the obstetrician may have to make himself aware of the accepted forms of early treatment for some of these conditions.

The readiness with which some deformities can be corrected if treatment is begun soon after birth and the increasing difficulty in obtaining correction if treatment is delayed is vitally important.

Manipulative treatments in for example, club foot and congenital dislocation of the hip have an infinitely greater chance of success if

begun at once. This success refers not only to the final results but prompt action greatly reduces the length of treatment with the attendent anxieties of parents. All this is dependant on the relaxation of structures seen at birth. A newborn is of course, very pliable and this property is retained for approximately the same length of time that other maternal hormonal influences remain evident. Many newborn babies can be refolded into the position that they occupied in the uterus. Such positions are of great variety and their reconstruction can sometimes explain club feet, torticollis and asymmetries as well as other secondary deformities. Intrauterine position may play some part in congenital hip dislocation for seventeen percent of a recent series of babies with congenital dislocation were presenting by the breech by comparison with a normal breech rate for the same department of four percent.

An unfortunately large proportion of congenital skeletal abnormalities are not amenable to any type of treatment but there are important exceptions.

Treatment is closely allied with a knowledge of the cause of congenital disabilities. We have mentioned the influence of intrauterine position on the foetus but this is only a mino r facet for familial, sexual, racial and geographic influences on the incidence of deformity are often important but only partially understood. One of the more striking examples of geographic variation is congenital dislocation of the hip which is virtually unknown in Singapore among the Chinese and Malay communities. In Japan however it is so frequent that it remains the major cause of degenerative arthritis of the hip in later life. Similarly the distribution of deformities may be seen to follow racial patterns even to the extent of suggesting common anthropoligical origins for racial communities.

Teratogenesis is a subject of great topical interset. Since the famous papers of Gregg in 1942 and 1945 drawing attention to the effects of maternal rubella in the first three months of pregnancy, it has become common knowledge that deformities may be produced by extraneous factors acting upon the embryo and this has more recently been thrust upon the public attention by the thalidomide disaster, These findings have of course, been widely followed up by an increasing range of experimental projects for, besides mechanical or genetic initiation of deformity, it is apparent that foetal abnormality may be caused by quite minor alterations in the enviromnent of the embryo. The time at which this interference takes place is more significant than the nature of the agent.

In the case of limb deformities it would seem that upset of the metabolic climate of the embryo on or about the fourth week of gestation may be responsible for the interruption of the normal processes of differentiation, development and growth and so may produce deformity. A great deal of attention has been directed on the evils of prescribing drugs to the pregnant woman in her first three months but this is far too simple for it is evident that teratogenesis can be induced before even the woman herself is aware of her pregnant state. This poses a problem when prescribing for any woman of childbearing age. The same consideration must be given by everyone to the hazards of ordinary X-ray examinations particularly of the pelvic and spinal areas for, during the first six weeks, the foetus is especially vulnerable to the effects of ionising radiation. If defects of the neural axis are due to toxic insults occurring in embryonic life then the neural tube is most vulnerable during the twenty-first to twenty-ninth day after fertilisation. That is, within seven to fourteen days of the first missed menstrual period.

It is important in this part of the World to discuss malnutrition as a teratogenetic factor for if the diet of the mother is poor she will in all probability produce a poor infant. Whenever environmental conditions or genetic conditions are unfavourable various manifestations of reproductive failure may be observed. At the extreme, sterility and the other manifestations such as high abortion rates, high still birth and neonatal death rates. The more severely deformed foetuses are therefore miscarried at an early date. Those congenital deformities which reach term—the true congential deformities—represent a relative reproductive success when compared with sterility or early embryonic death This explains why an increase in living standards —and this can be construed as an improvement of reproductive conditions in a biological sense

TABLE I

YEAR	CHINESE	MALAY	INDIAN	TOTAL
1961	47(47)	4(28)	0(0)	51(75)
1962	48(48)	6(42)	6(54)	60(144)
1963	42(42)	9(63)	11(99)	62(204)

will sometimes result in a higher incidence of congenital deformity.(Warkany 1947.)

Although overall figures are not great enough to draw significant conclusions, one might speculate from the figures for cleft palate and cleft lip in Singapore that this phenomenon is being witnessed in the Malay and Indian communities in Singapore (Table 1).

Table and absolute figures after Wong H.B.

The figures in parentheses allow correction for the relative proportions of births in the different communities.

For maternal dietary deficiency to result in congenitally deformed offspring, a nutritional borderline state must be created in the mother. Starvation and frank undernutrition lead to sterility and high peri-natal mortality. This dietary upset in the embryo is seen in diabetic women who present a high perinatal mortality and unhappily, produce above-average numbers of deformed babies.

The study of abnormalities inherent in the chromosomes can contribute towards the handling of genetically determined disorders in various ways. It is possible to advise affected carriers of recessive genes of the risk of transmitting the condition to their offspring or to advise parents on the likelihood of conceiving further handicapped children. Knowledge that a child is at risk may lead to early diagnosis and treatment; for example, in primary, dominant Vitamin-D resistant rickets, early therapy with large doses of Vitamin-D may avoid the onset of skeletal deformity. It is unfortunate that we must recognise that the genetics of many transmitted orthopaedic difficulties are not yet understood.

Often no single factor can be held responsible for defect and the final problem may be the result of permutations of racial, developmental and environmental factors possibly combining in different circumstances to produce a variety of skeletal abnormalities. There are a vast number of questions which remain without answer but in those cases where we are able to restore normal function or at least, do something to ease disablement, early appraisal and treatment are the foundations of success.

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