John Rendle-Short
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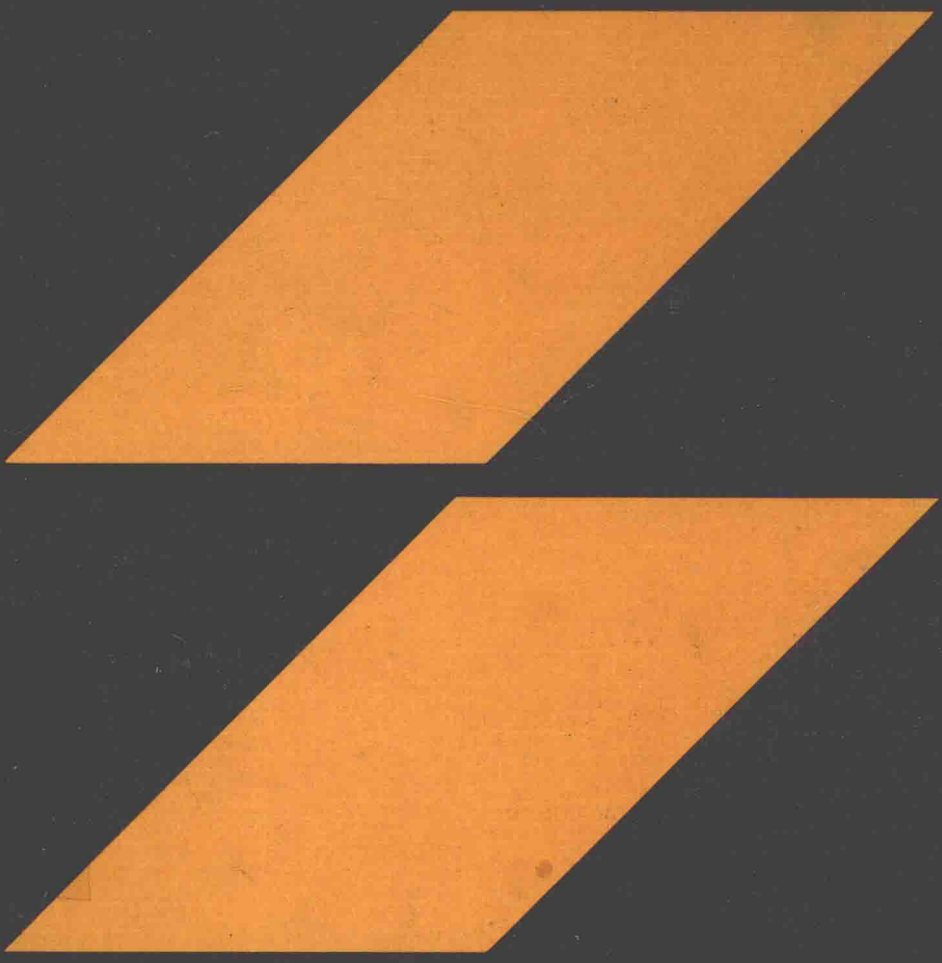
A synopsis of Children's Diseases

Sixth edition



WRIGHT

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Children's Diseases

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Preface to the Sixth Edition

This sixth edition of the Synopsis is smaller than the preceding one, and with an altered format is very nearly a new book. To bring a book up to date by the addition of much new material and simultaneously to reduce its size has proved a formidable task. It has only been possible because many diseases have been virtually eliminated by scientific progress. The huge area of social and psychiatric paediatrics, which occupies much of the modern paediatrician's time, has not been dealt with as it does not lend itself to the synoptic approach. Also the authors, who come from different geographical backgrounds, are well aware that social conditions differ from country to country. For these diseases, therefore, smaller, locally produced books are required.

The authors would like to thank Dr K. Verrier Jones for the chapter on 'Renal Diseases' and Doctors C. Wardrop and B. Holland for considerable help with the chapter on 'Blood Disorders'.

John Rendle-Short takes this opportunity to express his profound thanks to Peter Gray and John Dodge for their increasing help in the preparation of recent editions of the Synopsis, and to Messrs John Wright & Sons Ltd for their care in the production of six editions over 30 years.

J.R-S.

Brisbane, Australia

Preface to the First Edition

The importance of children's diseases is unquestioned; it was therefore considered that there was a place for a book of the synopsis type devoted to this speciality, in order to assemble all the relevant facts tidily for easy reference and rapid revision. It is hoped that this book will be of value to those preparing for examinations, whether Finals, the Diploma of Child Health or Membership of the Royal College of Physicians: also for Paediatric House Physicians and General Practitioners. It should not take the place of standard textbooks.

For the guidance of undergraduates, most diseases have been described as common, rare, etc. With few exceptions (e.g. rickets), those conditions marked as rare, or very rare, are of little importance to students taking their final examinations. It must be remembered, however, that whether a disease is described as common or rare depends largely on how many examples of that particular condition the author has seen.

The section on gastro-enteritis is purposely detailed, as the correct management of this disease is the key to the control of infantile dehydration and biochemical upset of whatever aetiology.

The appendix on drug dosage has been inserted especially for the benefit of House Physicians and General Practitioners. The doses have been checked by Mr A. Williams, PhC, MPS, Chief Pharmacist at Llandough Hospital, whose help is gratefully acknowledged. Dr T. Parry, Pathologist to Llandough Hospital, helped with the appendix on normal biochemical values.

Much of the material in this book is drawn from standard textbooks, especially Garrod, Batten and Thursfield's *Diseases of Children* and Mitchell-Nelson's *Textbook of Pediatrics* and grateful thanks are accorded to the writers of these and many other books and articles too numerous to mention individually. It is hoped that the authors will accept this as adequate acknowledgement. In a few instances, references have been given in a footnote, and illustrations taken from articles are acknowledged under each. Messrs Allen and Hanbury kindly supplied the illustration of the Woolwich nipple shield.

I would like to take this opportunity of thanking all those who read the proofs, and especially Professor A. G. Watkins, for much valuable criticism and advice. Several typists assisted me and my thanks are due to them, and particularly Miss W. R. Davies.

Finally to Messrs John Wright & Sons Ltd I express my sincere gratitude for their unfailing patience and help.

J R-S

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Section 1 Developmental Paediatrics

Chapter 1 NORMAL DEVELOPMENT

Developmental paediatrics is concerned with the maturational processes of normal and abnormal children from fetus to adulthood.

Growth refers to increase in size, anatomical and structural, measured in such parameters as height, weight, head circumference and bone age.

Development refers to increase in complexity in both structure and function. Its numerous simultaneous progressions are closely related but manifest many individual variations.

All children follow the same pattern of development—'they pass the same milestones', although the age at which they do so differs within certain limits, even in normal children.

Normally, growth and development of body, intellect and personality progress with age, and are fairly predictable in rate and outcome.

Abnormally, they are dissociated, producing widespread inconsistencies of and between somatic, cognitive and affective progressions, with unpredictable final results.

Factors affecting Growth and Development

Heredity determines the limits of each individual child's capacity to achieve optimal structural and functional maturity.

Environment determines the extent to which each individual child can fulfil his potential.

Purpose of developmental paediatrics is to monitor growth and development to:

1. Promote optimal physical, mental and emotional health.
2. Ensure early diagnosis and effective treatment of handicaps of body, mind and personality.
3. Discover the cause of any handicap.

A child's basic needs are for shelter and protective care; food; warmth and clothing; fresh air and sunlight; activity and rest; prevention of illness and injury; training in habits and skills necessary for the maintenance of life.

The psychological needs of both normal and handicapped children are for:

1. Dignity as a human being; different from an animal or machine; self-respect derived from knowledge of being valued as an individual.
2. Love and individual care.
3. Security based on (*a*) a sense of belonging, (*b*) stable interpersonal relationships, (*c*) familiar environment, but (*d*) not smothered with restrictions.
4. Discipline neither too severe nor too lax.

5. A sense of responsibility and opportunity to be of service to others, even though the child himself be handicapped.
6. Opportunity to achieve success in some field of endeavour.
7. Opportunity to learn from experience.
8. Opportunity to achieve personal, and if possible financial independence.

The Importance of Developmental Assessment

Understanding of normal childhood development with its many minor variations is essential for perceiving what is abnormal.

The normal developing child is active in body, intensely curious in mind and seeks a good relationship with adults and children.

Poorly developing children are inactive, not curious and have stunted personal relationships. If this is a chronic state, such children warrant observation and, if necessary, investigation.

1. If major delay occurs in all fields of development it may indicate:
   - 1.1 Prematurity—a pre-term baby at (say) 3 months post-natal age is not able to perform in the same way as full-term baby of the same age.
   - 1.2 Mental retardation.
   - 1.3 Cerebral palsy or other neurological or muscular disorder, e.g. amyotonia congenita.
   - 1.4 Severe illness causing weakness, e.g. gastro-enteritis.
   - 1.5 An emotionally deprived child.
   - 1.6 Sensory deficits, especially visual handicaps, but auditory and language difficulties also delay social behaviour and may lead to mistaken diagnosis of mental retardation.
   - 1.7 Infantile autism.
2. Delayed development of one area may occur in isolation, e.g.
   - 2.1 Delayed maturation. Sometimes a particular area of development (notably speech) fails to develop at usual rate. It eventually catches up and the child becomes completely normal in that area.
   - 2.2 Gross motor delay. Usually indicates physical disorder, e.g. dislocated hip preventing baby sitting.
   - 2.3 Fine motor delay. May indicate sensory loss or perhaps blindness.
   - 2.4 Delay in vocalizing or speech. May indicate deafness (*see* p.180).
   - 2.5 Minimal brain dysfunction (hyperactive syndrome).
   - 2.6 Specific learning difficulty in older child, dyslexia, etc.
3. Advanced development, occuring as an isolated phenomenon is of no prognostic significance for future intelligence, except cases of advanced speech development which may indicate high intelligence.
4. The greatest difficulty lies in the field of multiple handicaps. For example, it is very difficult to assess intelligence in a baby with athetosis and deafness who may nevertheless have normal intelligence.

Handicapped and Disadvantaged Children

A handicapped child is one who suffers from any continuing disability of body,

intellect or personality which is likely to interfere with his normal growth and development or capacity to learn.

Note: All handicapped children, however well provided otherwise, are disadvantaged and deprived in some way, and all disadvantaged and deprived children are handicapped socially even if not physically or intellectually.

A disadvantaged child is one who suffers from a continuing inadequacy of material, affectional, educational or social provisions, or who is subject to detrimental environmental stresses of any kind, which are likely to interfere with the growth and development of his body, intellect or personality, thus preventing him from achieving his inherent potential.

A handicapped child needs early identification of his disabilities and assets, prompt medical and surgical treatment and help and guidance for the parents to enable them to care for the child as long as possible in his own home. Appropriate training, education and vocational guidance and supervision and regular assessment throughout childhood and adolescence are also required. Finally, placement in the community or in special care.

The Child at Risk

The concept of the child at risk is of value as it enables the paediatrician to mark and follow children who have a poor prognosis. If, however, the net is cast too wide, too many children are followed and the exercise becomes unmanageable. Children at risk include those with an adverse family history, prenatal hazards, perinatal dangers, postnatal mishaps and developmental warning signals, e.g. the mother's suspicion that the child is not seeing, hearing, moving his limbs or taking notice like other children of his own age. She is usually right.

Other warning signals are: paediatric findings such as delayed motor development, lack of normal visual alertness, inattention to sound, delayed development of vocalization or speech, lack of interest in people or playthings and abnormal social behaviour of any sort. It is not safe to rely upon a single examination.

Comprehensive assessment of the handicapped child includes:

1. Neurological capabilities, sensory and motor.
2. Intellectual competence.
3. Social behaviour.
4. Paediatric examination, including careful evaluation of the child's visual and auditory capacity and his powers of communication.

Parent Guidance

1. Guidance regarding upbringing of a child to ensure optimum physical and mental health is required by all young parents. This is normally provided by grandparents in extended family. May be supplemented by paediatrician, books, etc.

2. Parents of handicapped children need additional support with knowledgeable instruction concerning everyday management. Special needs:

2.1 Truthful explanation of any handicapping condition, causation and prognosis.

2.2 Practical instruction in day-to-day care and management.

2.3 Continuing supportive counselling for all the family.

2.4 Referral to the appropriate social or medical agencies including provision of domestic help and financial assistance.

2.5 Realistic forward planning.
2.6 Genetic advice when necessary.

Management

1. Early training depends upon adequate stimulation. The natural teachers are the parents. The natural place is the ordinary family home. The natural tools of learning are playthings.

2. Education is an integral part of the handicapped child's treatment. Schooling must always be of prime consideration in planning medical and surgical procedures. Education may require special schools and facilities (e.g. school for deaf) but integration with normal children should be the aim as far as possible.

3. Vocational guidance must be based on realistic evaluation of the child's physical capacity, mental ability and social circumstances.

Ultimate Aims

'It is necessary for all concerned with the health, education and welfare of handicapped children to bear constantly in mind that childhood itself is a temporary phase in the life of any individual human being. The ultimate goal is to equip him or her in body, mind and personality to become, in adult life, a contented, self-reliant and useful member of the social community to which he or she belongs' (Mary D. Sheridan).

Patterns of Development

(For patterns of development from 4 weeks onwards, *see* Appendix 3).

The Baby at Birth

The baby at birth sleeps most of the time. When awake he is usually crying. He does not register pleasure. He dislikes a bright light shone into his eyes and responds by closing them. He turns towards a diffuse light, e.g. a window. Most of the time he lies immobile. He can flex and extend his legs and arms. When pulled into the sitting position his head falls back. When lying prone the infant cannot lift his head from the couch.

Jaw clonus is usually present, and occasionally ill-sustained ankle clonus.

The hands are held clenched, usually with the thumb between index and middle finger.

Primitive Reflexes. Many reflexes can be elicited in the newborn. Most are of academic interest only. Following may be of clinical importance as:

They are poorly developed or absent in pre-term or ill babies.

If they persist over age of 3 months, a neurological abnormality may be present.

Reflexes

1. Moro Reflex (age: birth–3 months) can be obtained by banging the cot-side, loud noise, etc. However, it is best elicited by holding the baby with the examiner's hand under the back, and if the head is then allowed to drop backward a few centimetres, the reflex should result:

Phase 1. Arms and legs are thrown out as if the baby were startled.

Phase 2. Arms are flexed as in embrace.

2. Grasp Reflex. Disappears at 6–8 months. This is elicited by examiner rubbing his fingers across the baby's palm. The baby grasps the finger firmly and can be lifted up by this means. It is strong in pre-term infants.
3. Asymmetrical Tonic Neck Reflex (ATNR) is elicited by turning the head to one side. The baby extends the arm and leg on that side and flexes on the opposite side. This fades at 36 weeks' gestation and is almost absent in full term, reappears at 1 month but later disappears.
4. Symmetrical Tonic Neck Reflex. Extensor tone in arms and flexor tone in legs when neck extended, reversed when neck flexed, disappears at 8–10 weeks.
5. Walk Reflex. Elicited by holding baby in standing position. Baby places one foot in front of other as though walking.
6. Step Reflex. If dorsum of baby's foot scraped along undersurface of table, he will step up onto table.
7. Rooting Reflex. If newborn baby's cheek is touched, he turns his mouth towards object which touched it. When baby is put to breast, therefore, and nipple touches cheek, baby turns mouth towards it.

Chapter 2 PHYSICAL GROWTH

Serial measurements of weight, height and head circumference are of great value in young children. They should be plotted on a centile chart (*Fig. 2.1*). Actual measurements are of less importance than whether or not serial measurements run parallel to the centiles.

Weight is more commonly used than height as a criterion of growth. Height, or length in an infant, is of equal importance to weight but considerably harder to measure and therefore less accurate. It has the advantage that a child cannot 'lose' height.

For prognosis of adult height, a bone age measurement (obtained by comparison of a radiograph of hand and wrist with accepted standards) is necessary.

Growth does not proceed regularly and uniformly. For the first 18 months of life there is a period of intense growth. From 18 months to 11 years growth occurs more slowly, about 5–6 cm/year.

At puberty there is a further period of active growth.

Tables of weight or height vary considerably according to the country of origin and date when they were compiled. For this reason local tables should be used as far as possible. Nevertheless international centile charts of value since it is the serial measurement which is important rather than spot checks, and they can also be used to relate weight and height to each other in assessment of nutritional state.

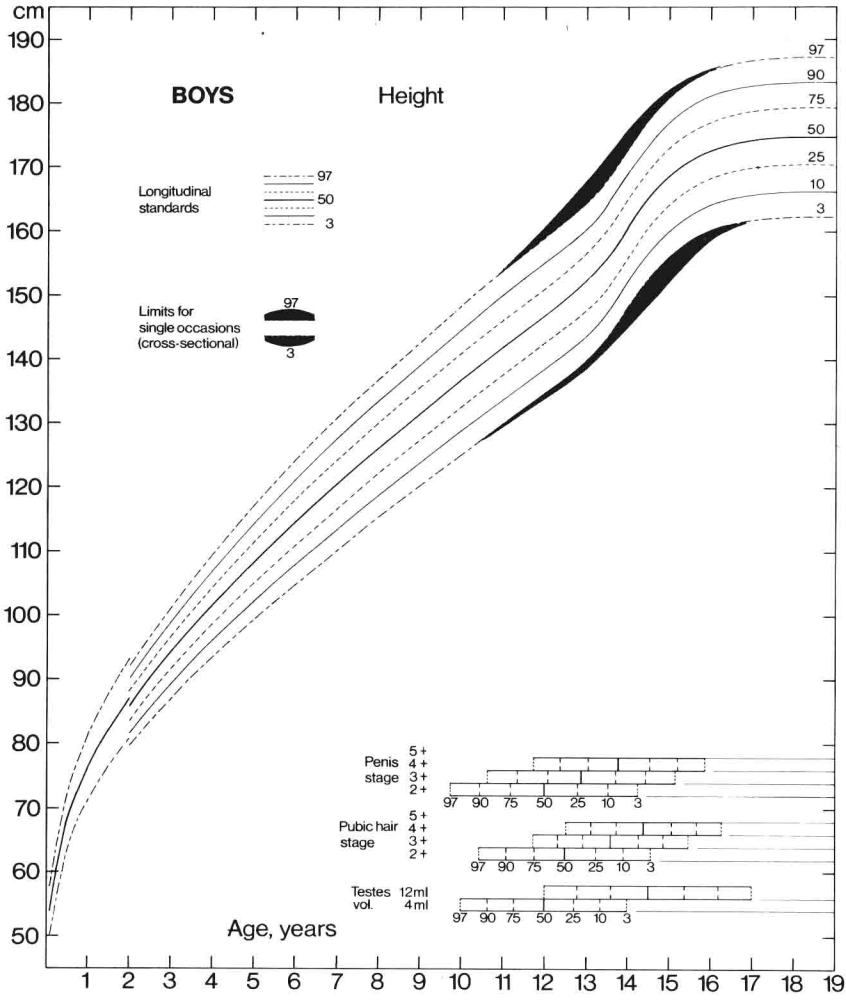
Some Factors influencing Physical Growth

1. Birth Weight

Children whose birth weight is low tend to remain relatively smaller than those with higher birth weight.

Fig. 2.1. Height chart for boys. (*By permission of J. M. Tanner and R. H. Whitehouse and Castlemead Publications, Hertford.*)

Name..... Date of Birth..... Reg. No. .....



2. Sex

Girls usually smaller than boys.

3. Familial Factors

Large parents tend to have large children.

4. Racial Factors

Some races, e.g. pygmies, tend to be shorter than others.

5. Endocrine Factors
These may influence weight or height: hypothalamic pituitary growth hormone, thyroxine, adrenal and gonadal hormones.

6. Environmental Factors
Children from low socio-economic groups tend to be smaller. All severe or chronic illnesses lead to a poor gain in weight or height.

Weight

The average weight at birth is 3400g (7½lb), but great variations occur. Weight of 4100g (9lb) not uncommon. If baby weighs 4600g (10lb) or more this may indicate that the mother is diabetic or pre-diabetic. The following factors can be used in calculating the expected weight in the first weeks of life: The baby normally loses weight after birth, but should have returned to his birth weight by the tenth day. He should gain approximately 30g (1oz) per day for the first 100 days of life. Expected weight can thus be calculated as follows: 30 × number of days since birth minus 10 equals weight in grams that child should have gained. This figure added to birth weight gives expected weight.

Fontanelles

The anterior fontanelle is normally closed by 18 months, but there is a wide range of normality. The posterior fontanelle should be closed by 2 months.

Teeth

Time of eruption varies greatly.

Eruption of Temporary (Milk) Teeth. Total number 20.
Lower central incisors: 6–10 months.
Upper central and lateral incisors: 8–12 months.
Lower lateral incisors and first molars: 12–18 months.
Canines: 18–24 months.
Second molars: 20–30 months.

Eruption of Permanent Teeth. Total number 32.
First molars: 6 years.
Central incisors: 7 years.
Lateral incisors: 8 years.

Puberty and Adolescence

Puberty occurs earlier in girls than boys. The physical sequence of events is given in Chapter 168.

Psychosocial adjustments in adolescence include achievement of identity and self-image, achievement of adult sexual role, achievement of independence from parents and family, and choice of career. Medically, adolescents are prone to emotional problems, physical trauma, infectious mononucleosis, venereal

disease, acne, obesity, anorexia nervosa and behavioural problems such as drug and alcohol abuse.

Chapter 3 MANAGEMENT OF THE NORMAL NEWBORN BABY

Immediate Management

1. Umbilical Cord

If conditions permit, the cord should not be clamped immediately. Allow placental transfusion for about 1 min, clamp and then do Apgar Score (*Table* 3.1).

Table 3.1. Evaluation of newborn baby

Apgar Score:* best scored 60s after birth of entire child and later at 5 min.
0=cardiac arrest and hence poor prognosis; 10=good prognosis.

Sign	*Score*		
	0	1	2
1. Heart rate	Absent	Slow (<100)	>100
2. Respiratory effort	Absent	Weak cry; hypoventilation	Good; strong cry
3. Muscle tone	Limp	Some flexion of extremities	Well flexed
4. Reflex irritability (response to stimulation of feet)	No response	Some movement	Cry
5. Colour	Blue; pale	Body pink; extremities blue	Completely pink

Note: Signs 1 and 2, greatest importance; 3 and 4, less; 5 least importance.
Acronym of Apgar Score:
A—appearance—colour
P—pulse—heart rate
G—grimace—reflex irritability
A—activity—muscle tone
R—respiratory—respiratory effort
See* Apgar V. (1953) Proposal for new method of evaluation of newborn infant. *Anesth. Analg.* (*Cleve.*) **32, 260; and Apgar V. (1966) The newborn (Apgar) scoring system: reflections and advice. *Pediatr. Clin. North Am.* **13**, 645.

2. Reception

Child should be received into warm, sterile towel, dried and kept under radiant heater if any doubt about condition.

3. Identification

Attach prepared arm/foot band with mother's name and number.

4. Vitamin K_1

Give 1·0 mg orally or intramuscularly.

5. Apgar Score at 1 and 5 minutes

If infant has Apgar score of 7 or less observe closely to determine need for resuscitation. Routine suction of oropharynx not necessary and may be harmful.

Later Management

Baby is observed in nursery for first few hours while mother recuperates. Observe respirations and colour, note whether baby 'mucusy'—if so pass catheter by mouth to check for oesophageal atresia.

Check umbilical cord for number of vessels, swab with 70% alcohol or apply triple dye or antibiotic spray.

Feeding

Considerable psychological help to mother if baby put to breast shortly after birth. Then infant goes to nursery until ready for a feed as judged by experienced nurse—usually within 6 h.

Medical Examination

Made as soon as is convenient after birth on all normal infants.

Objects

To find:

1. Any ill-effects from birth process.
2. Any congenital abnormalities.

Method

a. General Observation.

Look for:

1. Obvious congenital abnormalities, e.g. Down's syndrome (mongolism), spina bifida.
2. Colour
   - 2.1 Pallor (white)—a pale infant is a sick infant; pallor due either to shock or anaemia.
   - 2.2 Cyanosis (blue)—normal when confined to hands, feet, around mouth; abnormal if generalized and involves mucous membranes.
   - 2.3 Jaundice (yellow)—within first 24 h of life, is to be considered as due to haemolytic disease until proved otherwise.
   - 2.4 Plethora (red)—polycythaemia.
3. Posture
   - 3.1 Normal—infant lies on side; arms and legs flexed at side.
   - 3.2 Abnormal—infant lies on back; arms and legs abducted—frog position.
4. Spontaneous movements
   - 4.1 Normal—random. Asymmetrical. Note cry, and fine movements of face and individual fingers.
   - 4.2 Abnormal—jerky. Symmetrical. May be big movements. Or absence of movement.
5. Reactivity to stimuli
   - 5.1 In response to handling, undressing, pinprick, cotton wool stimulation of anterior nares.

5.2 In response to coincidental stimuli, e.g. door banging.

b. Top-to-toe Routine

Note: Routine examination can be by system, but for clinical convenience a good routine is to start at head and work down to toes; turn infant over and work back to head—the top-to-toe and back system.

1. Head
   - 1.1 Measure circumference.
   - 1.2 Palpate for swelling—caput or cephalhaematoma—or fracture.
   - 1.3 Feel fontanelles, especially tension, and over-riding of sutures.
   - 1.4 Note injury, e.g. from scalp electrode.
2. Face—capillary naevi over upper eyelids, central forehead, nose and upperlip—normal.
3. Eyes
   - 3.1 Notice any discharge—swab to laboratory if present.
   - 3.2 Pupil for coloboma—lens for cataract.
   - *Note*: Infant can be made to open eyes by giving a bottle to suck or holding upright.
4. Mouth
   - 4.1 Open by gentle pressure on mandible—look for cleft palate. Epstein's pearls on palate in midline—normal.
   - 4.2 If infant bubbly or history of maternal hydramnios—pass tube to exclude oesophageal atresia.
5. Ears—note accessory auricle, fistula or meatal atresia.
6. Jaw—micrognathos.
7. Neck
   - 7.1 Note: short in infants. Rotate and extend neck to:
   - 7.2 Feel sternomastoid for haematoma in lower part. Note branchial fistula. Feel clavicle for fracture.
8. Arms and hands—note full range of movements and correct number of digits.
9. Chest
   - 9.1 Respiratory rate, absence of insuction, coordinated breathing.
   - 9.2 Auscultation of little value—râles abnormal after first few hours.
   - 9.3 Note heart sounds and murmurs—feel apex beat. Pulsation in xiphoid region normal in first 48 h.
10. Abdomen
    - 10.1 Note umbilicus especially for infection or embryonic remains.
    - 10.2 Size of liver—normally 2 cm below costal margin.
    - 10.3 Feel for abnormal mass—kidneys are often felt normally.
11. External genitalia.
    - 11.1 Note any ambiguity of genitalia.
    - 11.2 Testes usually in scrotum.
    - 11.3 Vaginal skin tag frequent.

12. Hips—check for dislocation, *see* p. 440.
13. Femorals—palpate arteries—easier if legs flexed a little.
14. Legs and feet—note movements to be full; note posture of feet.

Turn infant over.

15. Inspect anus for patency, sphincter control.
16. Inspect and palpate back and neck—note dermal sinus or central naevus over spine.

c. Central Nervous System Evaluation

Already note made of posture, tone, reactivity, spontaneous movements.
Finally note reflexes:

1. Those normal in neonatal period: Moro; grasping reflex of fingers and toes; extension reflex of fingers and toes; rooting reflex; sucking and swallowing; lateral trunk incurvation; Galant reflex; walking reflex.
2. Tendon reflexes.

Multiple Births

Important to ascertain whether twins and sets of multiple births are monozygotic (identical) or dizygotic (not identical).

Criteria

1. Study of placenta—may give confusing results.
   1.1 Dizygotic twins have two separate placentae or single dichorionic placenta.
   1.2 Monozygotic twins usually have monochorionic placenta but may have separate placentae.
2. Sex, general appearance, hair colour, iris colour, iris pattern, blood group and serum protein pattern must *all* be identical for diagnosis of monozygosity. Environment and disease may modify height, weight and development (physical or mental).
3. Blood groups. Of factors listed in (2), blood groups are easiest to determine and most important. Following may be tested for: A–B, M–N, P. Lewis, Kell, Duffy and Rh.
4. Dermal patterns (dermatoglyphics). Extremely alike but never identical in monozygotic twins.

Note: Often possible to say categorically that twins are dizygotic but never possible to say categorically that they are monozygotic. Evidence will merely point to the fact that it is 99 per cent probable that they are monozygotic.

Prognosis

Second twin twice as likely to die as first. Probably from anoxia or cerebral trauma as second twin is often a breech. Aided delivery of second twin may be required.

Chapter 4 THE APPROACH TO CHILDREN

The growth of children, especially that of their brains, is greatest in infancy. Their emotional and cognitive functions develop most rapidly at this time. Adverse