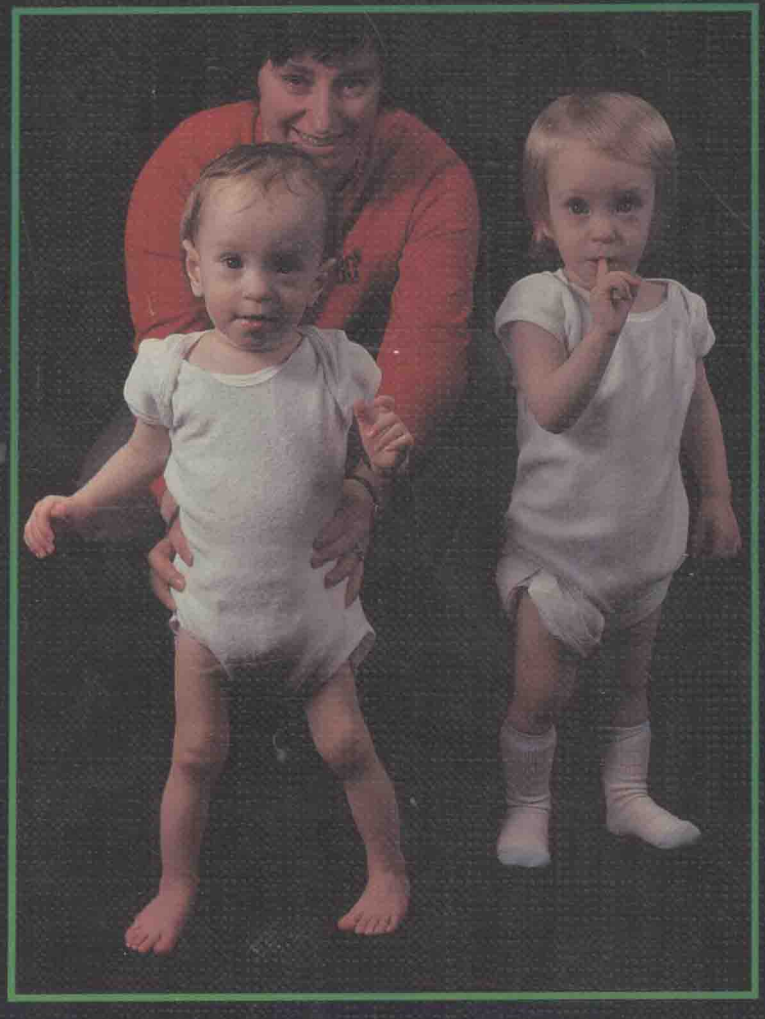
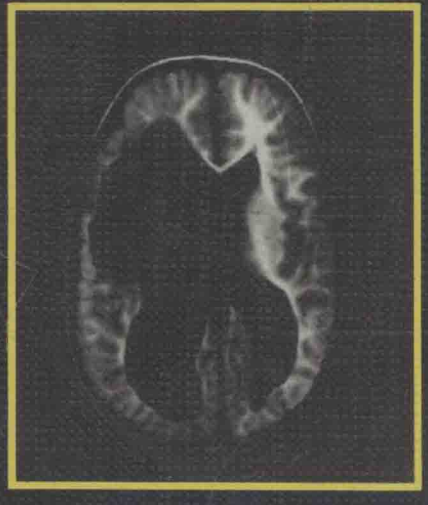
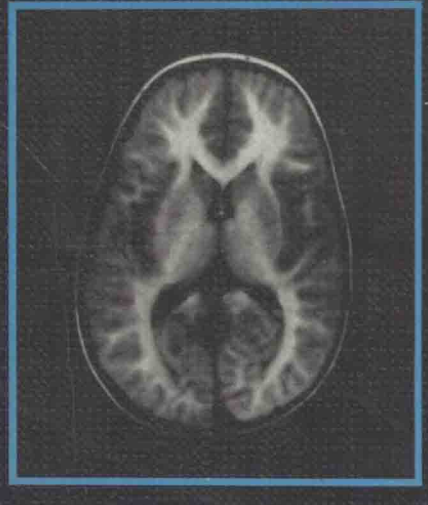
*Color Atlas of*

# Brain Disorders in the Newborn

*L.S. de Vries • L.M.S. Dubowitz*
*V. Dubowitz • J.M. Pennock*







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# Contents

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# Introduction

Haemorrhagic and ischaemic lesions of the brain during the perinatal period are a major cause of mortality and morbidity in the preterm and full term infant. The association between these lesions and the development of cerebral palsy has been recognised for several decades, but until recently these lesions could only be diagnosed at autopsy. The advent of X-ray computed tomography (CT) scanning and then ultrasound (US) imaging of the newborn brain made the diagnosis of brain lesions possible during life.

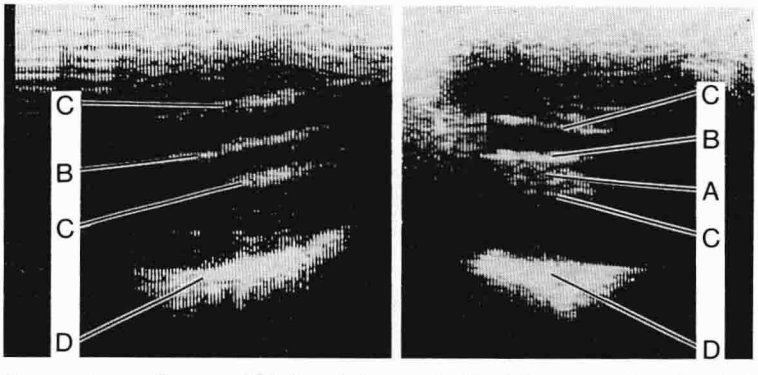
Haemorrhages in the brain were diagnosed in newborn infants by CT scanning in the late 1970s, but this technique was impractical for routine use as it involved the transfer of the infant from the neonatal unit to the scanner. On the other hand ultrasound equipment could be brought to the bedside; it was completely safe and non-invasive and repeated scanning could be done without any disturbance to the infant or interruption to her or his management. This opened the way not only for identifying haemorrhagic and ischaemic lesions of the newborn brain but also for following their evolution and resolution and correlating other modalities of assessment with the presence of these lesions.

The purpose of this atlas is to illustrate the way in which ultrasound, together with clinical assessment, electrophysiological studies, X-ray computed tomography and magnetic resonance imaging (MRI) has helped in the diagnosis, assessment and prognostication of these lesions.

## Ultrasound Imaging

The early linear array machines with 3.5 and 5 MHz transducers were capable of detecting haemorrhage which produced areas of increased echogenicity. The standard cuts were transverse (horizontal) through the temporal bone. On these images the presence or absence of haemorrhage could be identified (**1**).

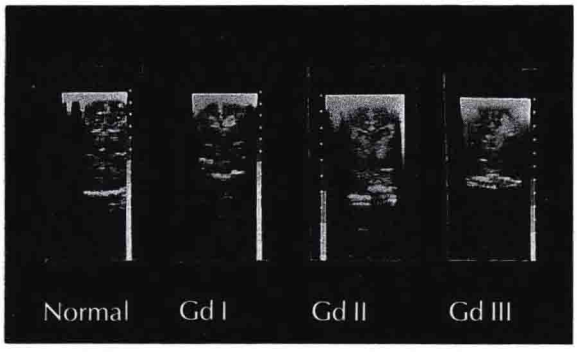
With improvement in the equipment, scanning through the anterior fontanelle became possible, allowing not only the diagnosis of haemorrhage but a better appreciation of the size of the lesion




1

Comparison of normal high axial scan (left) with a scan showing IVH in the lateral ventricle (A). Note the falx (B), lateral wall of lateral ventricle (C) and skull echo (D).

2

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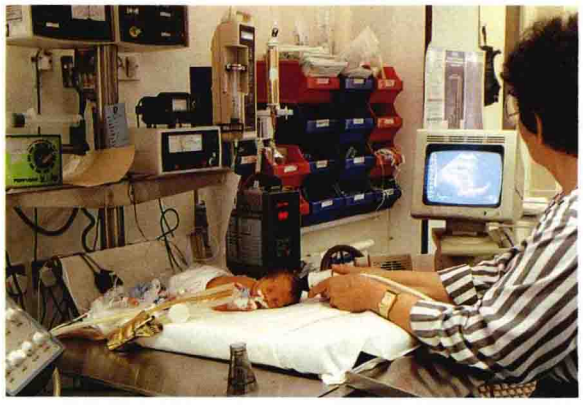
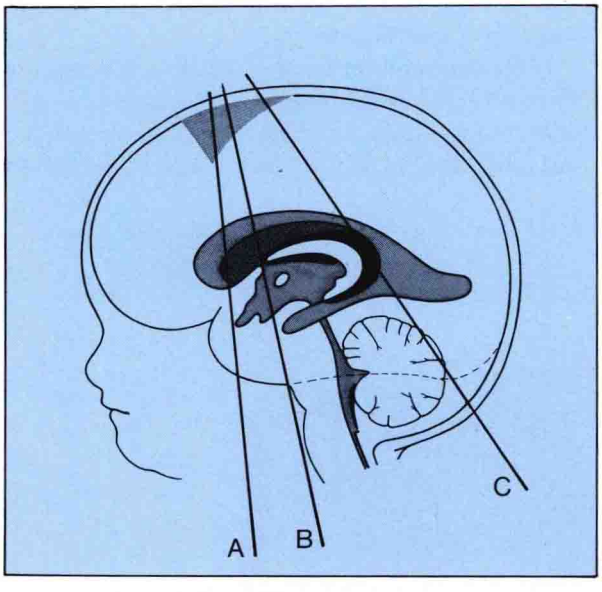


and of the lateral ventricles. However, the field visualised with the linear scanner was very narrow and the full extent of the lesions could not be appreciated (**2**)

The introduction of sector scanners with 5 and 7.5 MHz transducers (**3**) led to a marked improvement in resolution and also a wider field of view through the anterior fontanelle, with the ability not only to identify but also to grade lesions.

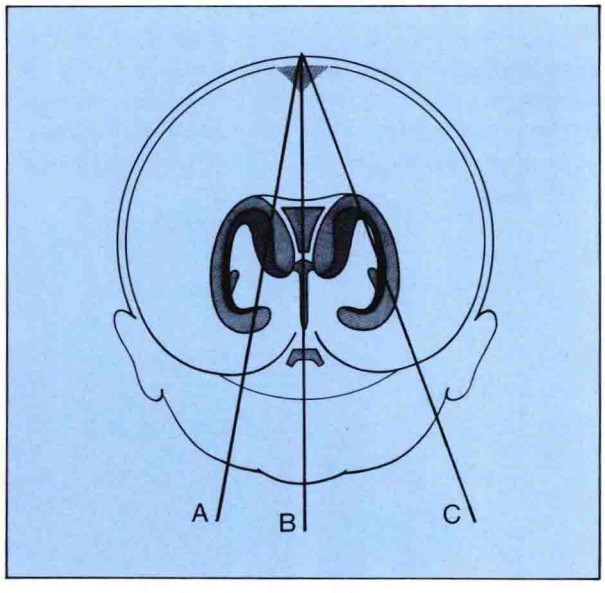
The angles of the standard cuts through the anterior fontanelle are shown in the mid-coronal plane, angling the scan head forwards through the frontal lobes and backwards towards the occipital lobes (**4**). Longitudinal sections in the sagittal and parasagittal planes parallel to the midline are shown in **5**.

4



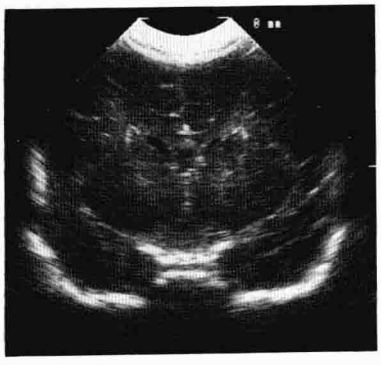

A = scan through frontal horn
B = scan through foramen of Monro
C = scan through trigone

5




A = scan through head of caudate nucleus
B = scan through the midline
C = scan through body of lateral ventricle

6



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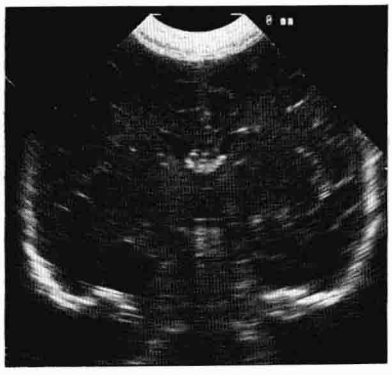

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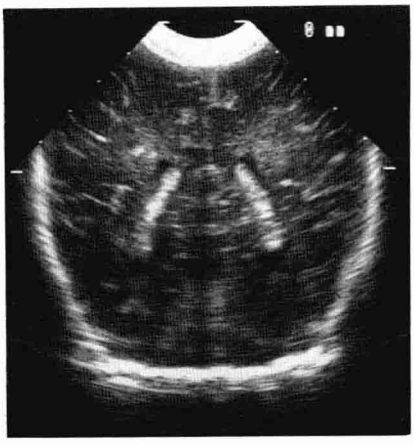

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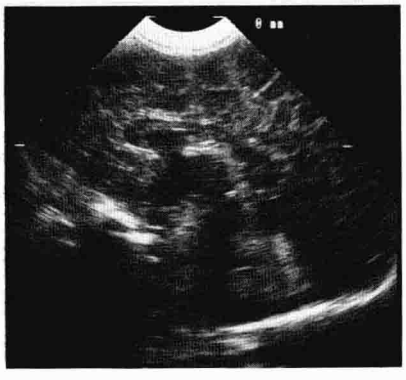

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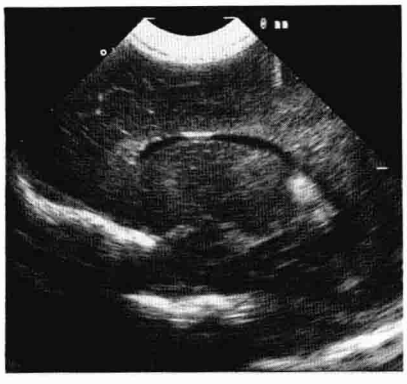
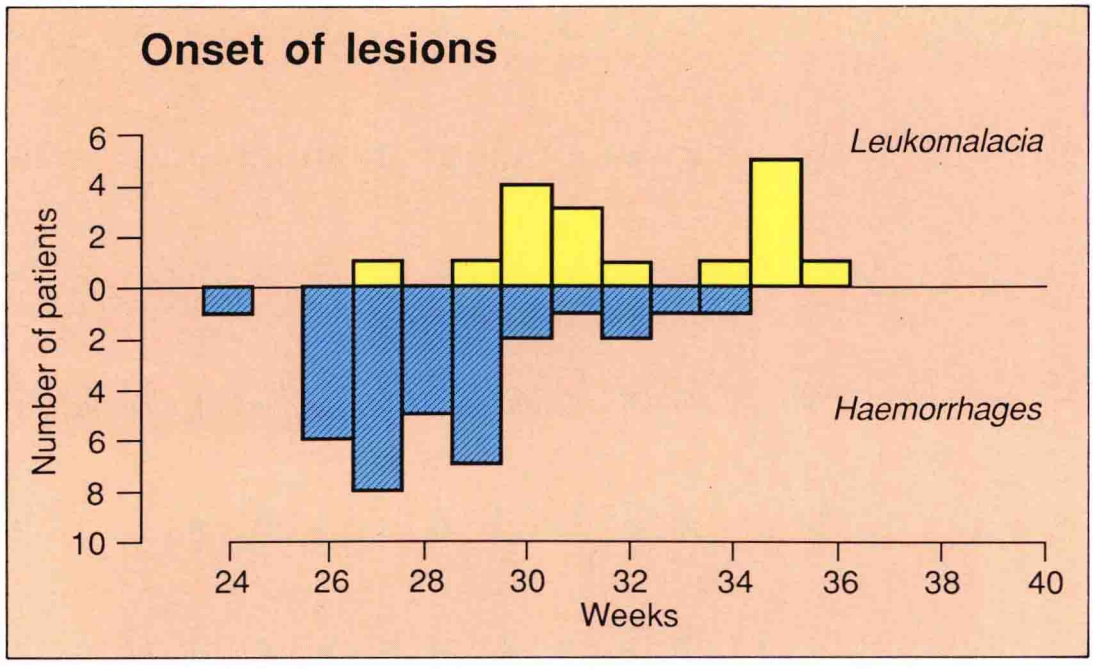


The normal appearances in the coronal cuts are shown in **6**, **7** and **8**, the sagittal and parasagittal views in **9** and **10**. The coronal ultrasound images are displayed with the right side of the child's brain on the right of the image. In the sagittal and parasagittal views the anterior horns are on the left of the picture and the posterior horns on the right.

With these standard sections it was possible to localise and grade haemorrhage accurately. Lesions in the periventricular areas such as periventricular leukomalacia, and lesions further out in the brain parenchyma, such as subcortical leukomalacia and infarction, could also be recognised.

Cumulative data on the occurrence and evolution of haemorrhagic and ischaemic lesions have produced useful information on the timing of these lesions during the postnatal period. Haemorrhages are rarely present at birth; most occur in the first 72 hours, and only a few after this period.

11




The relationship of different gestational age to haemorrhagic and ischaemic lesions can also be demonstrated (**11**). The value of repeated ultrasound scanning is summarised below.

**Value of Ultrasound**

- diagnosis of lesion
- type and site of lesion
- timing of lesion
- natural evolution
- response to treatment
- effect on clinical signs
- effect on physiological events

# Clinical Evaluation

When intraventricular haemorrhage was first recognised by Papile and her colleagues with CT scanning, they found a remarkably high incidence (approximately 50%) in preterm infants under 1500 grams. They noted that a large proportion of the infants appeared to be clinically 'silent' or normal. One possible explanation for this may have been the inadequacy of the clinical methods used for the evaluation of the nervous system of the preterm infant.

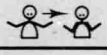
One of our main objectives was to evolve a system for the neurological assessment of the newborn infant which was sensitive, simple and reliable, as well as effective for the preterm and fullterm infant. It was also necessary to be able to monitor the progress of an individual infant and follow a large number of individual signs.

A system was evolved which integrated a selection of items reflecting posture, tone, primitive reflexes and neurobehavioural responses. These were tabulated in a proforma with a scoring system for individual signs of five grades, based on increasing response of that particular sign. Detailed instruction on the evaluation of each sign was also introduced in the proforma which could be used as a record for each individual examination. The documentation of individual items could be done directly on the proforma sheet by circling each individual sign (**12** and **13**).

By using a separate sheet for sequential examinations of the same infant, a comprehensive record of maturation in the normal 'optimal' premature infant, or the progress in an ill infant, could be documented. The evolution and resolution of deviant neurological signs could also be accurately and objectively recorded. Thus a comparison could be made of the neurological status of the preterm infant reaching 40 weeks postmenstrual age (PMA) with that of the newborn full term infant at 40 weeks gestation.

| NAME | D.O.B./TIME | WEIGHT | E.D.D. L.N.M.P. | E.D.D. U/snd. | STATES | STATE | COMMENT | ASYMMETRY |
|---|---|---|---|---|---|---|---|---|
| HOSP. NO. | DATE OF EXAM | HEIGHT | | | 1. Deep sleep, no movement, regular breathing.<br>2. Light sleep, eyes shut, some movement.<br>3. Dozing, eyes opening and closing.<br>4. Awake, eyes open, minimal movement.<br>5. Wide awake, vigorous movement.<br>6. Crying. | | | |
| RACE SEX | AGE | HEAD CIRC. | GESTATIONAL ASSESSMENT | SCORE WEEKS | | | | |

| | | | | | | | | |
|---|---|---|---|---|---|---|---|---|
| **HABITUATION** (≤state 3) | | | | | | | | |
| **LIGHT**<br>Repetitive flashlight stimuli (10) with 5 sec. gap.<br>Shutdown = 2 consecutive negative responses | No response | A. Blink response to first stimulus only.<br>B. Tonic blink response.<br>C. Variable response. | A. Shutdown of movement but blink persists 2-5 stimuli.<br>B. Complete shutdown 2-5 stimuli. | A. Shutdown of movement but blink persists 6-10 stimuli.<br>B. Complete shutdown 6-10 stimuli. | A. Equal response to 10 stimuli.<br>B. Infant comes to fully alert state.<br>C. Startles + major responses throughout. | | | |
| **RATTLE**<br>Repetitive stimuli (10) with 5 sec. gap. | No response | A. Slight movement to first stimulus.<br>B. Variable response. | Startle or movement 2-5 stimuli, then shutdown | Startle or movement 6-10 stimuli, then shutdown | A, B, C — Grading as above | | | |
| **MOVEMENT & TONE** | Undress infant | | | | | | | |
| **POSTURE** * (At rest — predominant) |  |  |  (hips abducted) |  (hips adducted) | Abnormal postures:<br>A. Opisthotonus.<br>B. Unusual leg extension.<br>C. Asymm. tonic neck reflex | | | |
| **ARM RECOIL**<br>Infant supine. Take both hands, extend parallel to the body; hold approx. 2 secs. and release. | No flexion within 5 sec.  | Partial flexion at elbow >100° within 4-5 sec.  | Arms flex at elbow to <100° within 2-3 sec.  | Sudden jerky flexion at elbow immediately after release to <60°  | Difficult to extend; arm snaps back forcefully | | | |
| **ARM TRACTION**<br>Infant supine; head midline; grasp wrist, slowly pull arm to vertical. Angle of arm scored and resistance noted at moment infant is initially lifted off and watched until shoulder off mattress. Do other arm. | Arm remains fully extended  | Weak flexion maintained only momentarily  | Arm flexed at elbow to 140° and maintained 5 sec.  | Arm flexed at approx. 100° and maintained  | Strong flexion of arm <100° and maintained  | | | |
| **LEG RECOIL**<br>First flex hips for 5 secs, then extend both legs of infant by traction on ankles; hold down on the bed for 2 secs. and release. | No flexion within 5 sec.  | Incomplete flexion of hips within 5 sec.  | Complete flexion within 5 sec.  | Instantaneous complete flexion  | Legs cannot be extended; snap back forcefully  | | | |
| **LEG TRACTION**<br>Infant supine. Grasp leg near ankle and slowly pull toward vertical until buttocks 1-2" off. Note resistance at knee and score angle. Do other leg. | No flexion  | Partial flexion, rapidly lost  | Knee flexion 140-160° and maintained  | Knee flexion 100-140° and maintained  | Strong resistance; flexion <100°  | | | |
| **POPLITEAL ANGLE**<br>Infant supine. Approximate knee and thigh to abdomen; extend leg by gentle pressure with index finger behind ankle. |  180-160° |  150-140° |  130-120° |  110-90° |  <90° | | | |
| **HEAD CONTROL (post. neck m.)**<br>Grasp infant by shoulders and raise to sitting position; allow head to fall forward; wait 30 sec. | No attempt to raise head  | Unsuccessful attempt to raise head upright  | Head raised smoothly to upright in 30 sec. but not maintained.  | Head raised smoothly to upright in 30 sec. and maintained  | Head cannot be flexed forward  | | | |
| **HEAD CONTROL (ant. neck m.)**<br>Allow head to fall backward as you hold shoulders; wait 30 secs. | Grading as above  | Grading as above  | Grading as above  | Grading as above  | | | | |
| **HEAD LAG** * Pull infant toward sitting posture by traction on both wrists. Also note arm flexion. |  |  |  |  |  | | | |
| **VENTRAL SUSPENSION** * Hold infant in ventral suspension; observe curvature of back, flexion of limbs and relation of head to trunk. |  |  |  |  |  | | | |
| **HEAD RAISING IN PRONE POSITION**<br>Infant in prone position with head in midline. | No response | Rolls head to one side | Weak effort to raise head and turns raised head to one side | Infant lifts head, nose and chin off | Strong prolonged head lifting | | | |
| **ARM RELEASE IN PRONE POSITION**<br>Head in midline. Infant in prone position; arms extended alongside body with palms up. | No effort | Some effort and wriggling | Flexion effort but neither wrist brought to nipple level | One or both wrists brought at least to nipple level without excessive body movement | Strong body movement with both wrists brought to face, or 'press-ups' | | | |
| **SPONTANEOUS BODY MOVEMENT**<br>during examination (supine).<br>If no spont. movement try to induce by cutaneous stimulation. | None or minimal<br>Induced | A. Sluggish.<br>B. Random, incoordinated.<br>C. Mainly stretching. | Smooth movements alternating with random, stretching, athetoid or jerky | Smooth alternating movements of arms and legs with medium speed and intensity | Mainly:<br>A. Jerky movement.<br>B. Athetoid movement.<br>C. Other abnormal movement. | | 1<br>2 | |
| **TREMORS**<br>Mark: Fast (>6/sec.) or Slow (<6/sec.) | No tremor | Tremors only in state 5-6 | Tremors only in sleep or after Moro and startles | Some tremors in state 4 | Tremulousness in all states | | | |
| **STARTLES** | No startles | Startles to sudden noise, Moro, bang on table only | Occasional spontaneous startle | 2-5 spontaneous startles | 6+ spontaneous startles | | | |
| **ABNORMAL MOVEMENT OR POSTURE** | No abnormal movement | A. Hands clenched but open intermittently.<br>B. Hands do not open with Moro. | A. Some mouthing movement.<br>B. Intermittent adducted thumb | A. Persistently adducted thumb.<br>B. Hands clenched all the time. | A. Continuous mouthing movement.<br>B. Abnormal toe posture.<br>C. Abnormal finger posture.<br>D. Convulsive movement. | | | |

| | | | | | | STATE | COMMENT | ASYMMETRY |
|---|---|---|---|---|---|---|---|---|
| **REFLEXES** | | | | | | | | |
| **TENDON REFLEXES**<br>Biceps jerk<br>Knee jerk<br>Ankle jerk | Absent | | Present | Exaggerated | Clonus | | | |
| **PALMAR GRASP**<br>Head in midline. Put index finger from ulnar side into hand and gently press palmar surface. Never touch dorsal side of hand. | Absent<br>R L | Short, weak flexion<br>R L | Medium strength and sustained flexion for several secs.<br>R L | Strong flexion; contraction spreads to forearm<br>R L | Very strong grasp. Infant easily lifts off couch<br>R L | | | |
| **PLANTAR GRASP**<br>Press the thumb against the ball of the infants foot. | No response<br>R L | Partial plantar flexion of toes.<br>R L | Toes curl around examiners finger.<br>R L | | | | | |
| **ROOTING**<br>Infant supine, head midline. Touch each corner of the mouth in turn (stroke laterally). | No response | A. Partial weak head turn but no mouth opening.<br>B. Mouth opening, no head turn. | Mouth opening on stimulated side with partial head turning | Full head turning with or without mouth opening | Mouth opening with very jerky head turning | | | |
| **SUCKING**<br>Infant supine; place index finger (pad towards palate) in infant's mouth; judge power of sucking movement after 5 sec. | No attempt | Weak sucking movement:<br>A. Regular.<br>B. Irregular. | Strong sucking movement, poor stripping:<br>A. Regular.<br>B. Irregular. | Strong regular sucking movement with continuing sequence of 5 movements. Good stripping. | Clenching but no regular sucking. | | | |
| **WALKING** (state 4, 5)<br>Hold infant upright, feet touching bed, neck held straight with fingers. | Absent | | Some effort but not continuous with both legs | At least 2 steps with both legs | A. Stork posture; no movement.<br>B. Automatic walking. | | | |
| **PLACING**<br>Lift infant in an upright position and allow dorsum of foot to touch protruding edge of a flat surface. | No response<br>R L | Dorsiflexion of ankle only<br>R L | Full placing response with flexion of hip and knee and placing sole of foot on surface.<br>R L | | | | | |
| **MORO**<br>One hand supports infant's head in midline, the other the back. Raise infant to 45° and when infant is relaxed let his head fall through 10°. Note if jerky. Repeat 3 times. | No response, or opening of hands only | Full abduction at the shoulder and extension of the arm<br>  | Full abduction but only delayed or partial adduction<br>  | Partial abduction at shoulder and extension of arms followed by smooth adduction<br>  <br>A. Abd>Add<br>B. Abd=Add<br>C. Abd<Add | A. No abduction or adduction; extension only.<br>B. Marked adduction only.<br>  | | J<br>S | |
| **NEUROBEHAVIOURAL ITEMS** | | | | | | | | |
| **EYE APPEARANCES** | Sunset sign<br>Nerve palsy | Transient nystagmus.<br>Strabismus. Some roving eye movement. | Does not open eyes | Normal conjugate eye movement | A. Persistent nystagmus.<br>B. Frequent roving movement<br>C. Frequent rapid blinks. | | | |
| **AUDITORY ORIENTATION** (state 3, 4)<br>To rattle. (Note presence of startle.) | A. No reaction.<br>B. Auditory startle but no true orientation. | Brightens and stills; may turn toward stimuli with eyes closed | Alerting and shifting of eyes; head may or may not turn to source | Alerting; prolonged head turns to stimulus; search with eyes | Turning and alerting to stimulus each time on both sides | | S | |
| **VISUAL ORIENTATION** (state 4)<br>To red woollen ball | Does not focus or follow stimulus | Stills; focuses on stimulus; may follow 30° jerkily; does not find stimulus again spontaneously | Follows 30-60° horizontally; may lose stimulus but finds it again. Brief vertical glance | Follows with eyes and head horizontally and to some extent vertically, with frowning | Sustained fixation; follows vertically, horizontally, and in circle | | | |
| **ALERTNESS/ RESPONSIVENESS**<br>Do not score appearance but responsiveness to visual stimulation. | Inattentive; rarely or never responds to direct stimulation | When alert, periods rather brief; rather variable response to orientation | When alert, alertness moderately sustained; may use stimulus to come to alert state | Sustained alertness; orientation frequent, reliable to visual stimuli. | Continuous alertness, which does not seem to tire, to visual stimuli. | | | |
| **DEFENSIVE REACTION**<br>A cloth or hand is placed over the infant's face to partially occlude the nasal airway. | No response | A. General quietening.<br>B. Non-specific activity with long latency. | Rooting; lateral neck turning; possibly neck stretching. | Swipes with arm | Swipes with arm with rather violent body movement | | | |
| **PEAK OF EXCITEMENT** | Low level arousal to all stimuli; never > state 3 | Infant reaches state 4-5 briefly but predominantly in lower states | Infant predominantly state 4 or 5; may reach state 6 after stimulation but returns spontaneously to lower state | Infant reaches state 6 but can be consoled relatively easily | A. Mainly state 6. Difficult to console, if at all.<br>B. Mainly state 4-5 but if reaches state 6 cannot be consoled. | | | |
| **IRRITABILITY** (states 3, 4, 5)<br>Aversive stimuli:<br>Uncover — Ventral susp.<br>Undress — Moro<br>Pull to sit — Walking reflex<br>Prone | No irritable crying to any of the stimuli | Cries to 1-2 stimuli | Cries to 3-4 stimuli | Cries to 5-6 stimuli | Cries to all stimuli | | | |
| **CONSOLABILITY** (state 6) | Never above state 5 during examination, therefore not needed | Consoling not needed. Consoles spontaneously | Consoled by talking, hand on belly or wrapping up | Consoled by picking up and holding; may need finger in mouth | Not consolable | | | |
| **CRY** | No cry at all | Only whimpering cry | Cries to stimuli but normal pitch | Lusty cry to offensive stimuli; normal pitch | High-pitched cry, often continuous | | | |

**NOTES** * If asymmetrical or atypical, draw in on nearest figure. Record any abnormal signs (e.g. facial palsy, contractures, etc.). Draw if possible.

**CHECK LIST OF ABNORMAL SIGNS**

| | |
|---|---|
| Head and trunk control | Orientation & alertness |
| Limb tone | Irritability |
| Motility | Consolability |
| Reflexes | Deviant sign |

Modified from *The Neurological Assessment of the Preterm and Full-term Newborn Infant*, by Lilly and Victor Dubowitz

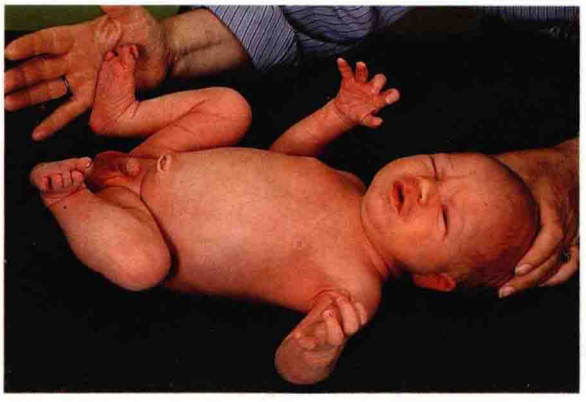


Record time after feed

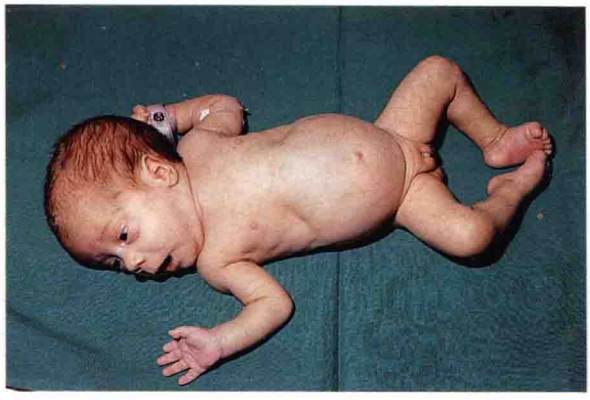
**EXAMINER:**

This comparison revealed a number of differences, including some in tone and posture. Differences between a full term infant during the first few days of life (**14**, **16** and **18**) and an infant born at 28 weeks gestation reaching 40 weeks PMA (**15**, **17** and **19**) are illustrated, in the supine position (**14** and **15**) and in ventral suspension (**16** and **17**). The difference in resistance to arm traction is illustrated in **18** and **19**. Thus it was possible to define a norm in preterm infants at 40 weeks postmenstrual age.

**14**

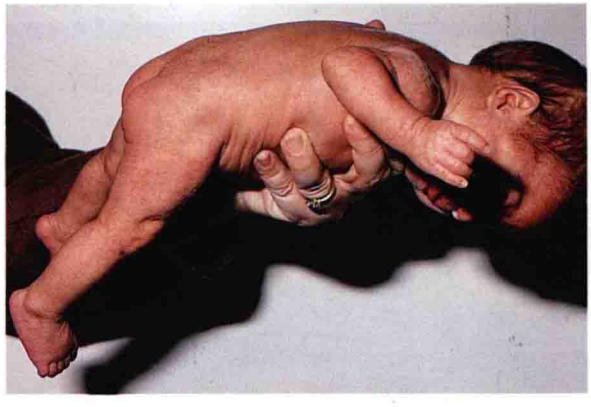


**15**



**16**

**17**



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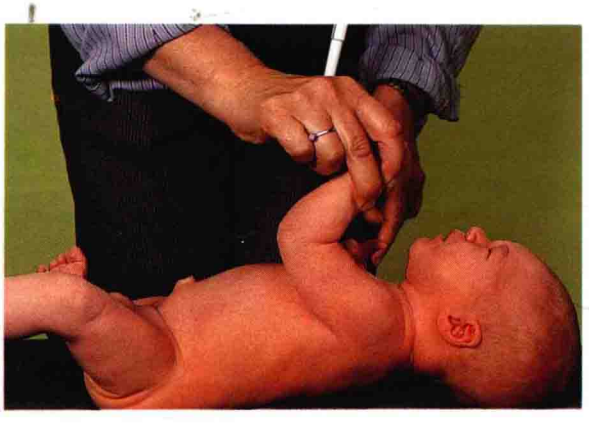

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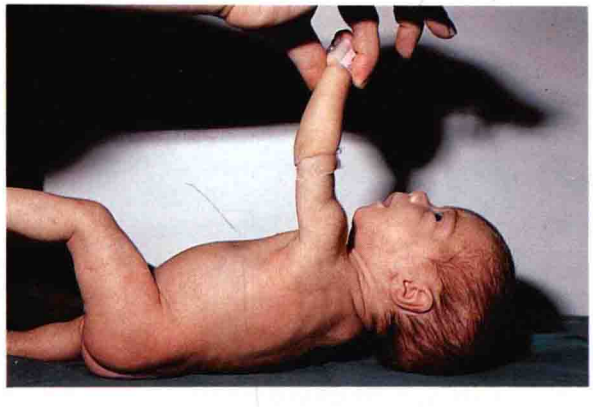
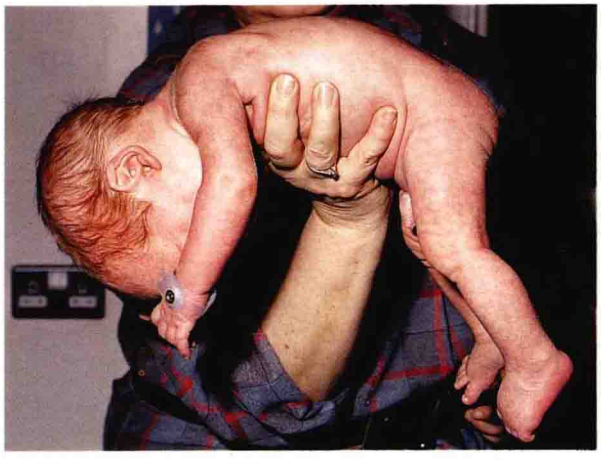


From this baseline, abnormal neurological signs in the individual ill infant could be recognised. These could take the form of abnormal maturation in relation to gestational age, which could be either delayed, 'accelerated' or deviant. Delayed development is illustrated in relation to development of poor posture of the head and trunk in ventral suspension (**20**), and poor head control when pulled to sit. Accelerated maturation may take the form of 'too good' head control in ventral suspension in an infant of 32 weeks PMA (**21**), due to excessive extensor tone.

**20**



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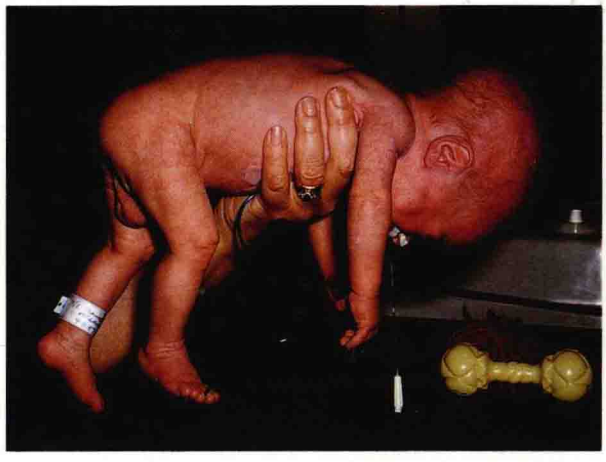

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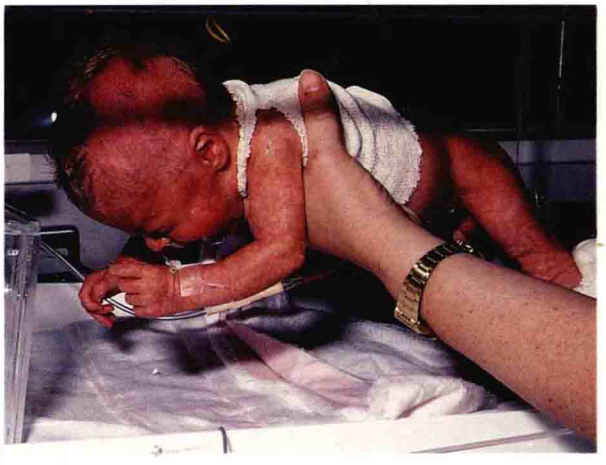

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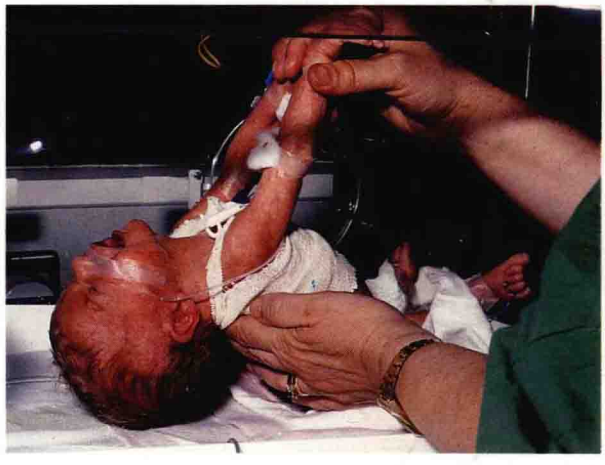


**Aberrant Signs**

- differential head control
- arm flexion > leg flexion
- tight popliteal angle
- frequent tremors and startles
- asymmetries
- absent plantar grasp
- abnormal Moro
- abnormal finger or toe posture
- irritability

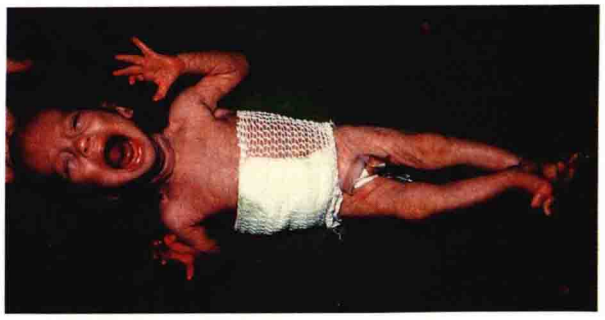
A number of aberrant signs can also be identified. Those of particular interest are summarised in the table (left).

Abnormal tone patterns showing differential head control are illustrated in **22** and **23**. Note increased extensor tone of the trunk and neck muscles in ventral suspension (**22**) and poor head control when pulled to sit (**23**).

Abnormal tone pattern consisting of marked flexor tone in the arm in association with increased extensor tone in the limbs is shown in **24**.

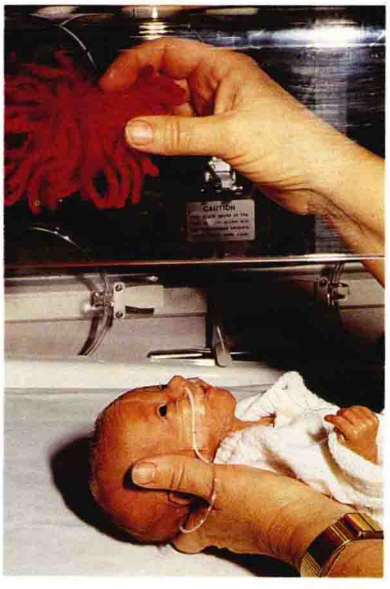
Also note the abnormal toe postures (spontaneous Babinski) and the abnormal finger posture, with flexion of thumb and index finger. The infant is also extremely irritable.

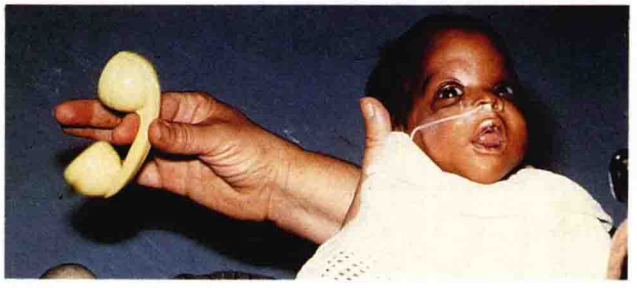
The detection of aberrant signs in individual infants and their correlation with the presence of haemorrhagic and ischaemic lesions on ultrasound examination have enabled us to recognise several



**24**

**25**





**26**

patterns of clinical signs. These are distinctive for a particular lesion and can be extremely useful in the clinical diagnosis of haemorrhagic and ischaemic lesions. Their persistence or/and resolution may also give a better basis for prognosis.

**Assessment of hearing and vision** forms part of the routine clinical examination and can be readily elicited by simple bedside testing. Visual tracking in the full term as well as preterm infant can be accurately assessed with a red woollen ball held at a distance of approximately 20cm (**25**).

Hearing is assessed with a simple plastic rattle which produces a white noise of 60–80 dB intensity. This is as sensitive as more sophisticated tools and will pick up both unilateral and bilateral hearing loss of 60 dB. The infant illustrated in **26** showed a brisk response on the left to the rattle but not on the right. Electrophysiological testing confirmed a hearing threshold of 80 dB on the left but a normal hearing threshold of 40 dB on the right.

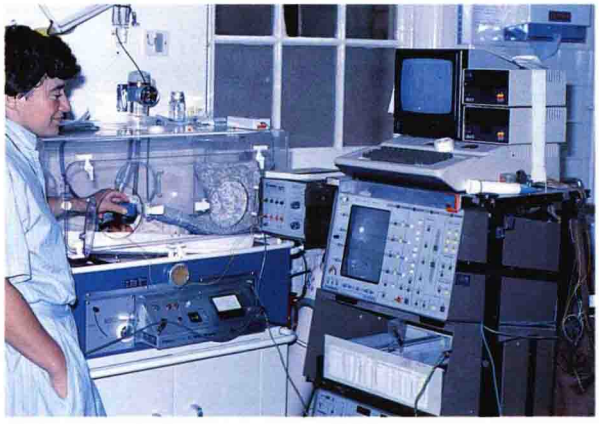
In addition to reflecting the state of the nervous system, visual and auditory responses may also help to pinpoint an isolated visual or auditory deficit. If suspected on clinical assessment, confirmation can be obtained by appropriate electrophysiological evaluation of the visual and auditory pathways.

# Electrophysiology

**Auditory Brainstem Evoked Responses (ABR)**: Measurement of ABR is a well standardised technique which is easy to perform in newborn infants (**27**). It involves providing an auditory stimulus via an earphone in the form of high frequency clicks, and recording the response via electrodes placed on the mastoid and vertex.

The fully mature response has six detectable waves (**28**), and information on different parts of the auditory pathway can be obtained by calculating the latency and amplitude of these waves and the intervals between them. The I–V latency is known as brainstem transmission time. **29** illustrates the maturation of these wave forms with gestational age. Note that latencies shorten and amplitude increases. ABR is of value in detecting both hearing difficulty and brainstem dysfunction.

27



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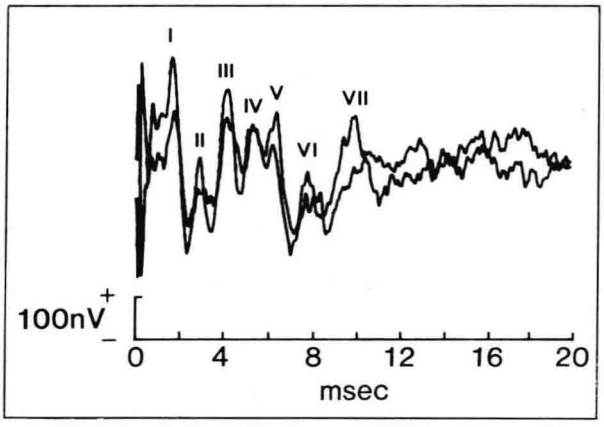


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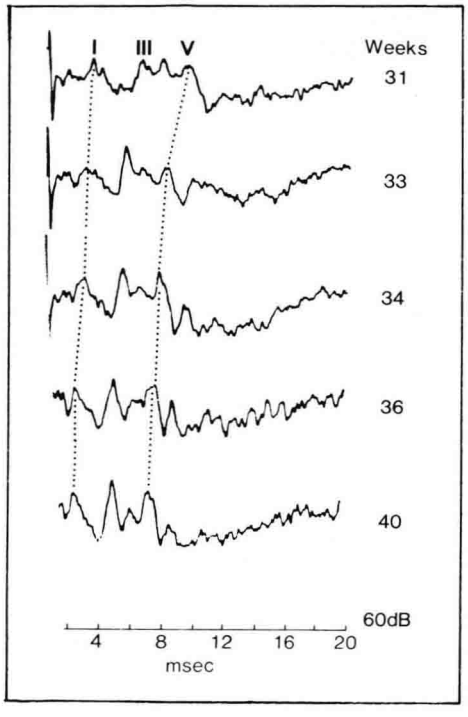


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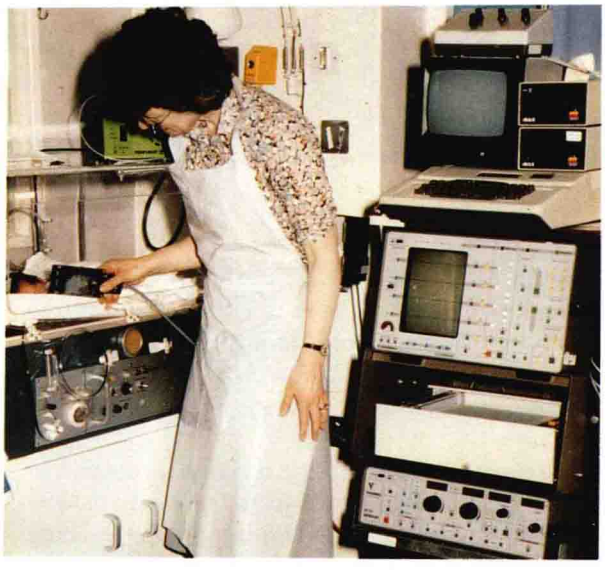
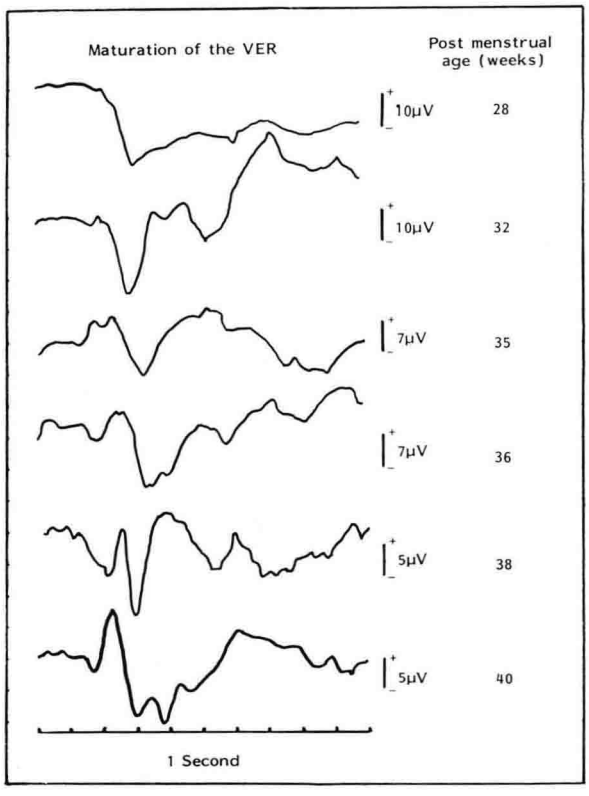


**Visual Evoked Responses (VER)** are obtained by using an appropriate visual stimulus such as a stroboscope or, preferably, diodes emitting red light (**30**). The latter can penetrate the closed eyelids of the infant, and a record of the response from the occipital cortex can be obtained. The mature trace shows a series of negative and positive waves.

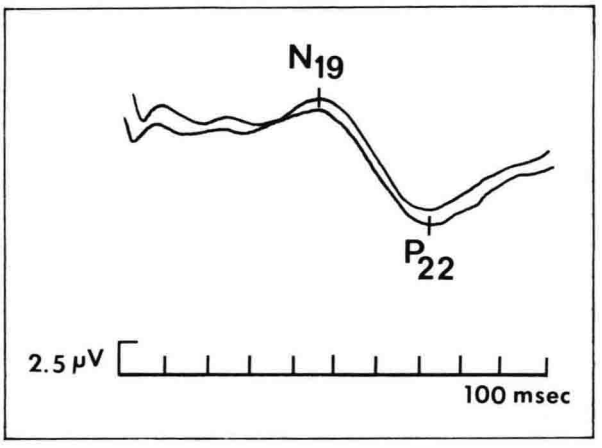
31




During early gestation the response consists of a negative wave only, but by 32–34 weeks a positive wave preceding the negative wave appears. The maturation of this response with gestational age (negativity recorded downwards) is given in **31**. VER is a sensitive test for the integrity of the visual pathways and the effect of lesions in the brain on these pathways. VER also plays an important part in verifying cortical blindness.

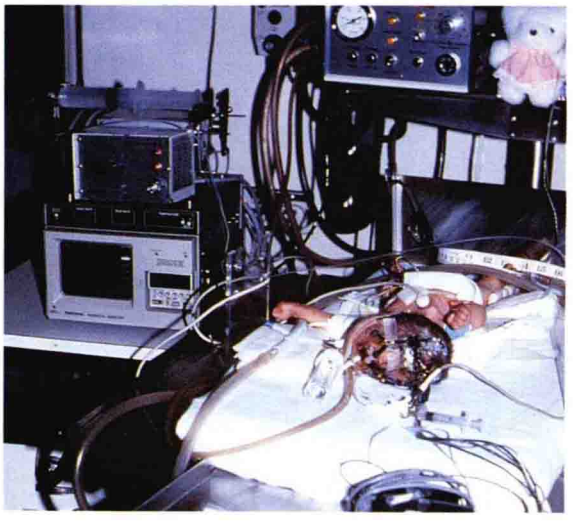
**Somatosensory Evoked Responses (SER)**: Following electrical stimulation of the median or tibial nerve, a response is recorded from the contralateral parietal cortex. It is of potential value in documenting the integrity of the sensory tracts of the nervous system. We have not used this technique routinely on our infants. Experience and data with this method are increasing, and SER have a strong predictive value for neurodevelopmental outcome. A normal recording is shown in **32**.

32




**Electroencephalography (EEG)**: With the introduction of the Oxford Medilog four-channel recorder it became practical to record continuous EEG activity in sick newborn infants without any distress to the infant or interruption to its intensive care or management (**33**). EEG recordings from 28 weeks gestation to term have been made to determine the process of maturation. Initially a very discontinuous pattern with intermittent bursts of activity is seen. This pattern becomes predominantly continuous, with only short periods of inactivity during quiet sleep, at term (**34**). By dividing 24 hour periods of recording into five minute epochs in optimal preterm and term infants, it has been possible to quantify the proportion of continuity and discontinuity of the tracings in relation to gestation, and to produce graphs of normal values (**35**).

33



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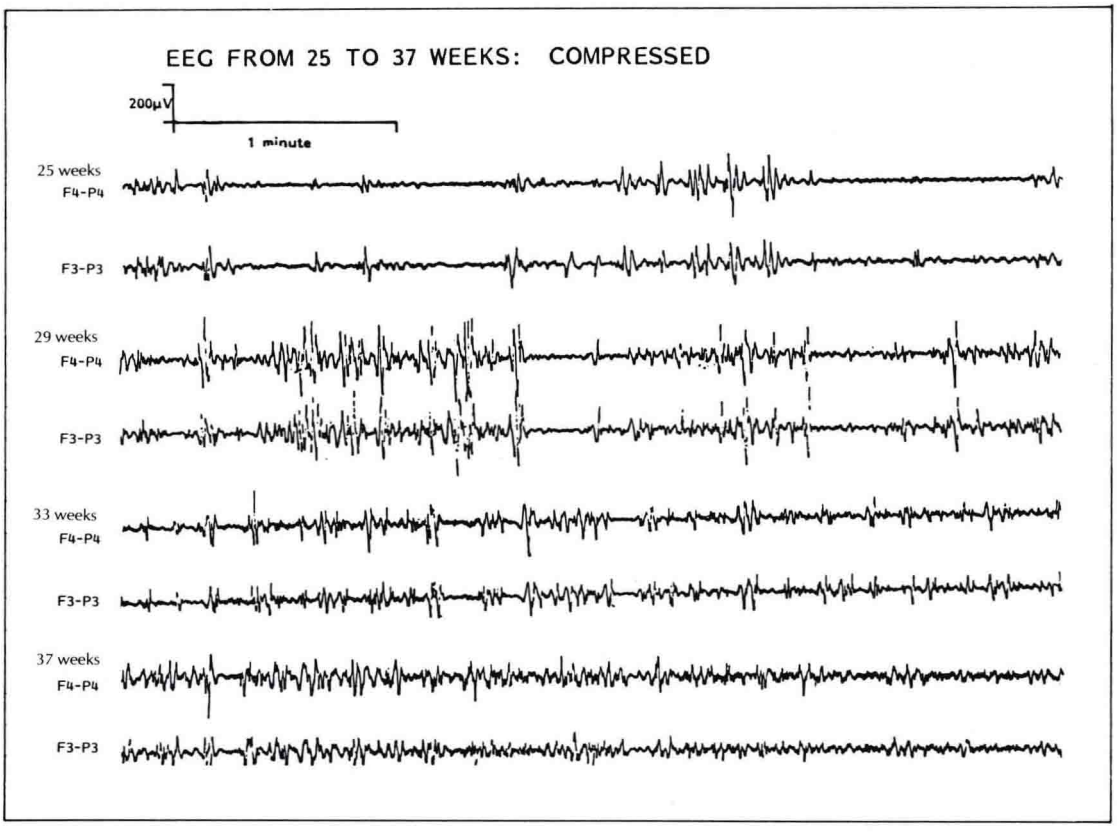


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