Acta Pædiatr 86: 757–61. 1997

The value of a scoring system for hypoxic ischaemic encephalopathy in predicting neurodevelopmental outcome

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Thompson CM, Puterman AS, Linley LL, Hann M, van der Elst CW, Molteno CD, Malan AF. The value of a scoring system for hypoxic ischaemic encephalopathy in predicting neurodevelopmental outcome. Acta Pædiatr 1997; 86: 757–61. Stockholm. ISSN 0803-5253

A numeric scoring system for the assessment of hypoxic ischaemic encephalopathy during the neonatal period was tested. The value of the score in predicting neurodevelopmental outcome at 1 y of age was assessed. Forty-five infants who developed hypoxic ischaemic encephalopathy after birth were studied prospectively. In addition to the hypoxic ischaemic encephalopathy score all but two infants had at least one cranial ultrasound examination. Thirty-five infants were evaluated at 12 months of age by full neurological examination and the Griffiths Scales of Mental Development. Five infants were assessed at an earlier stage, four who died before 6 months of age and one infant who was hospitalized at the time of the 12 month assessment. Twenty-three (58%) of the infants were normal and 17 (42%) were abnormal, 16 with cerebral palsy and one with developmental delay. The hypoxic ischaemic encephalopathy score was highly predictive for outcome. The best correlation with outcome was the peak score; a peak score of 15 or higher had a positive predictive value of 92% and a negative predictive value of 82% for abnormal outcome, with a sensitivity and specificity of 71% and 96%, respectively. For the clinician working in areas where sophisticated technology is unavailable this scoring system will be useful for assessment of infants with hypoxic ischaemic encephalopathy and for prognosis of neurodevelopmental outcome. □ *Cerebral palsy, hypoxic ischaemic encephalopathy, neurodevelopment, term infants*

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Perinatal asphyxia remains a major cause of mortality and neurodevelopmental disability in term infants. In developing countries the incidence of postasphyxial neurological damage is particularly high (1, 2). The clinical neurological sequelae in the immediate neonatal period following perinatal asphyxia are referred to as hypoxic ischaemic encephalopathy (HIE). These sequelae have been shown to be better predictors of outcome than Apgars scores and blood gases (3).

Hypoxic ischaemic encephalopathy was originally described by Amiel-Tison in 1969 (4) and there have been numerous studies since then. Recently, new technologies have become available to determine cerebral damage more accurately and earlier in the perinatal course. These technologies include computed tomography (CT) scanning, magnetic resonance imaging (MRI), cerebral function monitoring, cranial ultrasound and doppler ultrasound of the middle cerebral artery (5–8). These modalities are, however, not available in many neonatal units, and certainly not in developing countries. There is a need for a simple but accurate clinical method of predicting outcome.

The most widely used classification of HIE is that of Sarnat and Sarnat (9), which groups affected infants into one of three categories: mild, moderate and severe. The decision as to whether an infant falls into the moderate or severe category is at times difficult and the outcome of infants in the moderate group is variable. Application of this grading system is also time consuming and requires some paediatric expertise.

More recently, three published studies have developed scores for HIE. Portman et al. (10) developed a score that predicts early morbidity and mortality. Two other papers developed scores which have been related to long-term outcome. One (11) used the postasphyxia score and the other a neonatal behavioural neurological assessment (12). These latter scores are lengthy, involving 17 or more measures, and require some training. Both, however, have found value in the use of such scores in predicting neurodevelopmental outcome.

We have put together a scoring system at Groote Schuur Hospital's Neonatal Intensive Care Unit which is numeric with fewer items. It is based on that of Sarnat and Sarnat but is much simpler and we hope that it will fill the need in the developing world for an unsophisticated, yet predictive assessment of infants with HIE. This paper reports on the evaluation of this scoring system in terms of its predictive value for neurodevelopmental outcome at 1 y of age.

The HIE score

The score consists of a clinical assessment of nine signs (Table 1).

Table 1. Hypoxic ischaemic encephalopathy score.

Sign	Score 0	1	2	3	Day 1	Day 2	Day 3
Tone	Normal	Hyper	Hypo	Flaccid			
LOC	Normal	Hyper alert, stare	Lethargic	Comatose			
Fits	None	Infreq $< 3\ d^{-1}$	Frequent>2/day				
Posture	Normal	Fisting, cycling	Strong, distal flexion	Decerebrate			
Moro	Normal	Partial	Absent				
Grasp	Normal	Poor	Absent				
Suck	Normal	Poor	Absent ± bites				
Resp.	Normal	Hypervent	Brief apnoea	IPPV (apnoea)			
Font'l	Normal	Full, not tense	Tense				
				Total score per day			

Tone

The tone progresses from normal and slightly increased peripheral tone in the mildly affected infant to the more severely affected infant who is generally hypotonic or completely flaccid.

LOC (level of consciousness)

The assessment of LOC is as described originally by Sarnat and Sarnat (9). The mildly affected infant has a normal LOC or is hyperalert and staring with normal or decreased spontaneous movement and exaggerated responses to minimal stimuli. The more severely affected infant progresses through lethargy to complete unresponsiveness ("stuporose", as described by Sarnat).

Fits (clinically apparent seizures)

The score increases with increasing frequency of seizures.

Posture

This is assessed again as described by Sarnat and Sarnat (9) but in this study an intermediate score of 1 is given to the infant who has mild to moderate HIE and who shows intermittent bicycling movements of the limbs together with fisting (thumbs flexed, adducted and opposed across the palms).

Moro, grasp, suck (the primitive reflexes: moro reflex, palmar grasp and suck reflex)

These reflexes are normal in the mildly affected infant, poor or partial in moderate HIE and absent in severe HIE.

Resp (respiratory pattern)

In mild HIE the infant breathes normally or hyperventilates. More severely affected infants have episodes of apnoea and may require ventilation.

Font'l (fontanelle tension)

The more severely affected infant may have a full or tense (bulging) fontanelle.

Each sign is scored from 0 to 3 and the score for each day is totalled. The higher the score the more severely affected the infant. The maximum possible score on any one day is 22. The score is equally applicable in a ventilated infant. It cannot be applied in a paralysed infant.

Patients and methods

Term infants of 37 weeks' gestation or more (as determined by the Ballard score) were selected for the study if clinical signs of HIE developed after birth.

Infants were scored by attending intensive-care physicians daily until the score was 0 or until hospital discharge. Ten infants were subjected to scoring independently by two observers over a period of 4 d, yielding 40 paired scores. An interobserver reliability coefficient of 0.87 was obtained.

The only objective investigation generally available to us was cranial ultrasound. At least one cranial ultrasound examination was carried out, usually prior to hospital discharge. Infants were followed up at 18 weeks and 1 y of age.

An infant neurological assessment was conducted at 18 weeks of age, and at 12 months of age the Griffiths Scales of Mental Development (13) and a full neurological examination were performed by a paediatrician blinded to the infant's history.

Outcome was considered abnormal if there was clinical evidence of cerebral palsy, defined as a nonprogressive motor disability, or developmental retardation, defined as a Griffiths' general quotient less than 70.

Results

Forty-five infants entered into the study over a 10 month period. The outcome of 40 (89%) infants was known at 1 y of age, and five infants were lost to follow-up.

Table 2. Characteristics of the mothers and their infants.

	Number ($N = 40$)	%
Mothers:		
Mean age, y (SD)	24 (6)	
Black race	32	80
Primiparity	23	53
Teenagers	13	29
NVD	25	63
Caesarean	6	15
Instrument	7	18
Infants:		
Birthweight, g (SD)	3375 (502)	
Range	2400–4950	
Male	27	68
1 min APGAR < 6	33	83
5 min APGAR < 6	20	50
Mean cord pH (SD)	7.11 (0.12)	
Mean cord BD (SD)	14.21 (5.02)	
Seizures	33	83
Ultrasound:		
Normal	17	43
Oedema	11	28
SCL	9	23
HIE score:		
Max peak score < 11	14	35
Max peak score > 15	13	33
Normal day 7	13	32

Profile of mothers (Table 2)

Fifty-three per cent of the mothers were primigravidae and 29% were teenagers. Sixty-three per cent delivered vaginally, 18% with instrumentation and 15% by Caesarean section.

Profile of infants (Table 2)

Two-thirds of the infants were male. Fifty-eight per cent were delivered at a tertiary hospital. Thirty-three (83%) had a 1 min APGAR score less than 6 and the same proportion had seizures. Only one-third of all the infants became neurologically normal by d 7 (i.e. scored 0).

Thirty-eight infants had a cranial ultrasound (the other two infants had mild HIE and were discharged early): 21 infants (56%) had an abnormal cranial ultrasound. Of these abnormal scans, nine showed subcortical leucomalacia (SCL). One infant had a small intraventricular haemorrhage (not shown in Table 2).

Thirteen (33%) infants had maximum HIE scores of more than 15, 14 (35%) 10 or less and the remainder (13 or 33%) between 11 and 15. The cohort was therefore evenly spread over the range of the score.

Outcome

Twenty-three infants were normal (58%), 16 (40%) had cerebral palsy with varying degrees of developmental retardation and one infant was developmentally delayed (hypotonic with motor delay but functioning normally in all other areas). Four of the 16 infants with cerebral palsy died in hospital before 6 months of age.

A Griffiths Developmental Assessment was performed on 35 infants at 1 y of age (the other infant was hospitalized with cerebral palsy and uncontrolled seizures at the time of assessment). The overall mean general quotient was 92 (range 7–128). The neurologically abnormal infants had a mean GQ of 47 (range 7–101) and the normal infants 116 (range 94–128). There was a significant negative correlation between the GQ and the peak HIE score, with a correlation coefficient of 0.7.

The mean daily scores and 95% confidence intervals of the normal and abnormal infants over the first 10 d of life are shown in Fig. 1. There was a significant difference throughout ($p = 0.03$ on d 1 and $p < 0.01$ thereafter).

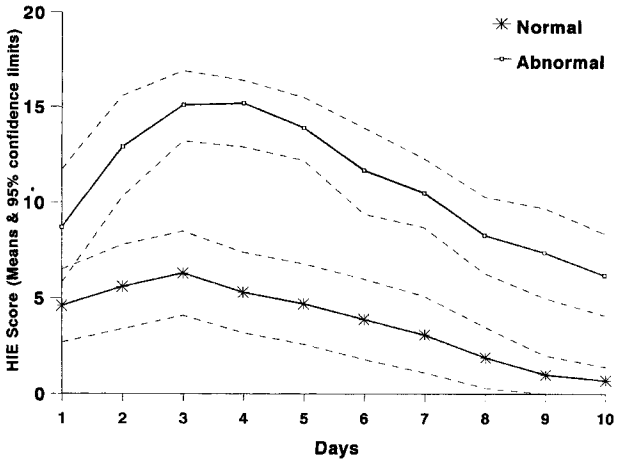

Fig. 1. HIE mean score: profiles of normal and abnormal infants.

Predictive values (Table 3)

Predictive values for abnormal outcome both for the score itself and for clinical findings associated with HIE were calculated (Table 3).

The presence of clinical seizures had a positive predictive value (PPV) of 57% for abnormal outcome but a

Table 3. Predictive values, sensitivity and specificity of the HIE score for abnormal outcome at 1 y of age.

	PPV (%)	NPV (%)	Sensitivity (%)	Specificity (%)
Seizures	57	92	94	48
Leucomalacia	100	74	53	100
Max score > 10	65	100	100	61
Score d 3 > 10	73	94	94	74
Score d 4 > 10	75	90	88	78
Max score > 15	92	82	71	96
Score d 3 > 15	89	71	47	96
Score d 4 >15	90	73	53	96
Abnormal score d 7	63	100	100	57
Abnormal d 7 and max score > 15	92	100	100	93

negative predictive value (NPV) of 92%. An infant with HIE who did not have seizures had a 92% chance of being normal. However, the presence of SCL had a PPV for abnormal outcome of 100%. All of the infants seen at 1 y who had SCL on ultrasound had cerebral palsy (seven spastic quadriplegics and two microcephalic with hypotonic cerebral palsy). The NPV was 74% (some infants without SCL were abnormal).

Analysis of the HIE score also revealed some useful trends. All infants whose peak HIE score was 10 or less were normal at 1 y of age (i.e. the NPV of a maximum score greater than 10 was 100%). Sixty-five per cent of infants with a peak score above 10 were abnormal, whereas 92% with a peak above 15 were abnormal. Seventy per cent of infants with a d 3 or 4 score of more than 10 were abnormal (PPV 73% and 75%, respectively), whereas 90% of infants with a score of more than 15 on these days were abnormal.

All those infants whose score had returned to 0 by d 7 were normal (the NPV was 100% for an abnormal score on d 7). When assessed in combination, an infant with a peak score above 15 and who remained abnormal after d 7 had a 92% chance of being abnormal. All infants who peaked below 16 and who were normal by d 7 were normal (NPV 100%).

The infants were retrospectively classified into Sarnat and Sarnat (9) categories by folder perusal by clinicians blinded to the outcome of the infants. The oculovestibular and tonic neck reflexes as well as the autonomic function parameters and electroencephalographic assessment were excluded. The results are shown in Table 4. In this table the numerator of all fractions indicates the number of abnormal infants and the denominator indicates the total number of infants in the group. The majority of damaged infants were in the severe group. All infants in the mild group were normal. There were three cerebral palsy infants in the moderate group and the infant with developmental delay also fell into this category. In general, infants in the mild group had an HIE peak score of 10 or less, the moderate group 11–14 and the severe group more than 14. There were three infants whose HIE score fell outside these ranges. One infant in the severe group peaked at 14 and two infants in the moderate group peaked at 17.

Table 4. Number of infants with cerebral palsy related to Sarnat grading and maximum HIE score.

	Maximum HIE score	Sarnat Mild	Sarnat Moderate	Sarnat Severe
0/10*	0–10	0/10	0/0	0/0
3/13*	11–14	0/0	2/12	1/1
14/17*	15–22	0/0	2/2	12/15
		0/10**	4/14**	13/16**

*Number of infants with cerebral palsy/total number of infants in HIE score category.

**Number of infants with cerebral palsy/total number of infants in Sarnat category.

Discussion

It is evident from recent research that HIE in the long term produces a spectrum of neurological disabilities and impairments (14, 15). Newer sophisticated diagnostic modalities will become more accurate in predicting these disabilities as research progresses, but these technologies are unlikely to become available in the developing world. What is needed in this setting is a simple clinical tool which will reliably predict the infant's outcome so that families can be provided with information as early as possible and plans for intervention for the disabled child can be implemented.

We have found two other studies that have used a clinical grading system. The postasphyxia score (PAS) of Lipper et al. (11) has 17 items and the neonatal behavioural neurological assessment of Bao et al. (12) has 20 items. In the latter paper the authors state that a 2 week training course is required before the clinician can use the assessment. It is not clear whether specific training for the PAS was instituted. Both methods appear to predict neurological outcome adequately early on in the perinatal course—that of Lipper et al. was predictive even as early as 24 h.

Robertson and Finer (15) have found that the prognostic value of the stage of encephalopathy is greatest when the newborn neurological examination is graded according to the most severe signs and evaluated over a 7 d period. We propose that our grading system, which incorporates some items from both previously published scores and takes into account both the most severe signs and the length of persistence of these signs, is as accurate, is quicker to administer, requires no additional training for medical and paramedical personnel and requires no equipment. It is ideal for implementation at any district or rural facility where special investigations and indeed specialist paediatricians are unavailable. It contains many of the features included in the three stages of Sarnat and Sarnat but excludes autonomic function, the deep reflexes and some of the primitive reflexes. We have added the grasp reflex, the respiratory pattern and a clinical assessment of the fontanelle tension since ultrasound is not generally available in developing countries.

Ultrasound data were reported as, locally, this investigation was readily available. These data were incomplete in that two of the 40 infants were not scanned, but both infants were normal at their 1 y assessment. Seventy-five per cent of scans were done after d 7. Of the 10 scans done before d 7 all the infants were normal at 1 y of age. Eleven infants had evidence of oedema on ultrasound, four of whom had cerebral palsy, and it is possible that late SCL was missed. If this was so it would only add weight to the predictive values of SCL on ultrasound and would not affect the other results.

In keeping with other developing countries (2), severe asphyxia is common in our population. The cause is not clear but may be a combination of an inadequate pelvis and a large infant. Table 2 shows that there is a relatively low rate of Caesarean section, which may affect the severity of the cases included in this cohort. There was a high

incidence of SCL in this cohort. It must, however, be appreciated that this is not a prevalence study but a study of the usefulness of a clinical assessment. Therefore the whole spectrum of HIE is represented.

We use predictive values in evaluating the usefulness of neurological assessments, as do other workers in this field (16, 17). Predictive values indicate the likelihood of disability and are thus more meaningful than sensitivity and specificity, which express retrospective relationships (18).

There was a peak in mean score values on d 3 and 4 (Fig. 1). The predictive values of the score were therefore determined on these days (Table 3) as well as for d 7 and for the individual maximum score. The maximum score for each individual infant yielded a higher predictive value than that for d 3 or 4.

Evaluation of the infant on d 7, as found by other authors, is a useful time with regard to prognosis (19). Sarnat and Sarnat also indicated that normality after d 6 automatically placed an infant in the mild and therefore low-risk group (9). Our highest predictive values were at this time-point.

So far, this cohort has only been followed to 1 y of age, at which time it is difficult to assess mild mental retardation; however, a cut-off DQ of 70 was taken. Outcome was described in terms of normality or abnormality. One infant, who had a suspect neurological examination and a normal DQ, was included in the normal group for the purpose of analysis.

The predictive values of the score are high, especially when the peak score and length of persistence of abnormal signs are considered together. Predictive values of 92% and 100% and sensitivities and specificities of 100% and 93%, respectively, are excellent and are higher than those of CT scanning in the study by Lipper et al. (11).

Robertson and Finer have also defined perinatal variables useful for predicting good outcome in the moderate encephalopathy group and suggest that infants in this category can be discharged at an early age from follow-up. We propose using our score to categorize infants into a low-risk category, thus allowing for health-care workers working distant from tertiary medical centres to decide which infants can be discharged safely from their overcrowded clinics. Use of the graphical representation of the cohort (Fig. 1) may allow the rural clinician to superimpose the patient's profile and thereby predict outcome or allow discharge from follow-up.

In conclusion, the HIE scoring system that we have adapted for our needs and which has become an integral part of the care of the encephalopathic infant in our service: (i) is easy to use and can be adequately applied by junior staff members; (ii) correlates with the Sarnat and Sarnat descriptive grading; and (iii) has a high predictive value for outcome.

Certainly, the parents of an asphyxiated infant who scores a maximum of 10 or less and is normal by d 7 can be assured of a normal outcome. Those infants whose score peaks higher than 15 and who remain abnormal after d 7 must have a guarded prognosis.

Acknowledgments.—The authors gratefully acknowledge the invaluable contribution of Dr Sandro Buccimazza who performed some of the Griffiths assessments, Mrs Vivien Magasiner who helped with the 18-week assessments and evaluation of the infants at 1 y of age who had cerebral palsy, and Mrs Priscilla Baleni, whose tireless searching for defaulters ensured that the follow-up rate was achieved. We also thank the mothers and their infants who often travelled long distances to attend follow-up visits—their efforts, we hope, will ensure better care for those who follow. This project was funded by Mobil.

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Received Aug. 2, 1996. Accepted in revised form Feb. 28, 1997