INFANTILE FUNDUS LESIONS IN RELATION TO MENTAL CAPACITY*

BY

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Developmental abnormalities and other congenital defects of the eye often remain unsuspected in the early weeks or months of life because newborn babies do not as a rule undergo detailed intraocular examination. For research purposes a clinic may go out of its way to scrutinize the ocular fundi and media in every infant, but in ordinary out-patient and office practice the need seldom arises. We know that the macula is not fully developed at birth, even in healthy full-term children, so that nurses, parents, and others in close contact with infants do not usually feel perturbed about a baby's blank stare—unless that vacant look persists beyond the time when a healthy baby should be taking in his entourage with a purposive stare. If later on we discover a bilateral macular coloboma or any other fundus lesions ample to account for defective sight we can as a rule tell the parents that nobody could have prevented the trouble, and that no treatment, however promptly it had been given, could have made the eyes normal.

Before mentioning some of the diseases in which fundus lesions may give a clue to the mental state, let us consider what kind of situation makes a parent or doctor wish to have an infant's eyes examined.

1. Ophthalmoscopy may be done as a routine precaution in the absence of any unfavourable signs. Perhaps there is a history of eye trouble in other members of the family, or perhaps the parents are full of fear through having often heard eye defects discussed.

2. An explanation may be sought for abnormal signs in the eyes. For instance, one eye may be smaller than the other, or possibly there is nystagmus, proptosis, squint, inequality or peculiar shape of the pupils, or opacity of one or both lenses.

3. The victims of certain disease-states are prone to suffer from ocular complications, so that ophthalmoscopy may serve to clinch the diagnosis, as in Tay-Sachs disease or in toxoplasmosis. Then again, the family doctor may seek to know whether the optic disks are atrophic in cases of hydrocephalus, oxycephaly, or other deformities of the skull and face.

4. There is the important group of infants who make their mothers anxious because they do not gaze concentratedly at things, and fail to follow objects with their eyes by the time they are expected to do so. In the early stages suspicion is more likely to settle upon the faculty of sight, although in fact the blank expression often turns out to be a sign of mental defect; and this brings us to the important question whether we can make a reasonable forecast of the future intelligence by observing patients during infancy.

Early Recognition of Mental Defect

Medical opinion is far from unanimous concerning the chances of early assessment. Kirman (1953) said: "It is only very seldom that a child of less than 2 can safely be diagnosed as definitely mentally defective." His view

agreed with that of Bowlby (1951), who wrote: "Mental tests have no predictive value in the first 18 months of life." Gesell's (1943) attitude was radically different from that of Kirman and Bowlby. He came to the conclusion that nearly all cases of mental deficiency can be diagnosed in the first year of life—except those which arise from secondary causes in later infancy.

Does the truth lie midway between these extremes? I think not. In my opinion Gesell's conclusion is far more realistic. It is, of course, true that from time to time the mind of an apparently defective infant will develop far better than at one stage we could have hoped, because medicine is far from being an exact science. Every experienced physician, and many a layman as well, can cite instances of a patient being given a few weeks more to live, and then perhaps recovering from the allegedly fatal illness and enjoying rude health a year or two later. These happy turns of fate are some compensation for all the suffering which we meet in everyday practice, but they do not alter the general prognosis by more than a small fraction. The same principle applies to infants' intelligence. Early asphyxia or some other form of birth trauma can hold up a baby's progress, so that he may for a while seem defective and yet later catch up; but in most backward infants the stigmata are not only evident but also lasting.

Here it must be emphasized that Gesell's view was based upon a vast amount of clinical material. He and his colleagues examined in detail and made follow-up observations on many thousands of infants and children. We cannot shut our eyes to the fact that uniform backwardness during the first year of life nearly always means permanent defect of mind. Backwardness in a few isolated aspects may be manifest without there being any lowering of the general level, but *general* backwardness is nearly always ominous.

Criteria of Mental Development

At the Great Ormond Street Hospital for Sick Children a few years ago I was wading through some not easily legible notes about a child who had been referred to me, and I found myself asking: "What are these *millstones* he keeps talking about? *Must* the children have them?" "Oh, sir," said the resident registrar, or deputy clinical supervisor of lower age groups, or whatever the modern title is for what we used to call H.P. (house-physician), "it isn't *millstones*, it's *milestones* and it's development." "What's development?" I replied, somewhat pedantically, I fear; "but," I went on, "never mind. Development is religion, and I don't want to interfere with that." "Time which antiquates antiquities," to borrow Thomas Browne's expression, has also accustomed and reconciled me to this usage of "milestones," which now seems to be one of the favourite words of paediatricians.

These milestones or *stages* in normal development form a useful series of criteria, so long as they are not too rigidly interpreted as a measure of the infantile mind. For instance, a mentally alert baby's failure to pass this or that milestone can be due to blindness, muscular weakness, or skeletal defect. Therefore a milestone test is valid only when it is reckoned in the light of all available facts about the baby's physical state. With this precaution in mind, and remembering also that milestones are merely a series of averages or approximations, we are entitled to regard each milestone as a useful gauge; and it is now generally agreed that in former times too much attention was paid to the dates of tooth-eruption and closure of the anterior fontanelle as yardsticks of infantile progress.

Unfortunately we cannot always obtain accurate information about the previous milestones, because parents are often unreliable when they come to recount their own child's progress. A fond mother can see a smile flit across her baby's heavenly countenance when in fact he is only being blown into facial spasm by wind bombinating around his abdomen. Incidentally the time-table of development as recounted by the father may wildly conflict with what the mother has

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worked out. Even so, we should never omit to take a careful history when we try to estimate an infant's mental level.

An average healthy baby of 2 weeks can momentarily hold up his head when he is held sitting. At 4 weeks he should be able to look fixedly at his mother when she talks to him, and a smiling response to her blandishments is likely to dawn when he reaches the age of 6 weeks. By this time he should be able for a few seconds to lift up his head in the line of his trunk when he is held in the prone position by a hand placed under his abdomen. At 3 months he should be able to turn his head in the direction of a sound that interests him, and at this stage he can usually clutch an object placed in his hand. At 6 months or slightly sooner he should be able to crawl a little distance towards a desired object, and to sit unsupported in a pram, though probably another month will elapse before he can sit without support on a flat surface like the floor. If his hands are held at 10 months he can, after a fashion, walk (or at least do a rock 'n' roll), and launch into a solo stagger by the end of his fifteenth month. Overt drooling usually finishes at the age of 1 year, although many non-certifiable persons continue, figuratively speaking, to drool for the rest of their lives.

Persistence of a blank stare well beyond the age of 6 weeks usually makes the mother anxious, but in many instances it is some outside person, often the doctor, who first suspects abnormality. Naturally the lack of expression upon an infant's face will be all the more striking to someone who has not been watching him every day. At first the mother is unlikely to suspect any mental trouble unless the baby was born with some gross defect or has been attacked by a severe convulsive illness. When she comes to realize that by this time her baby should have been watching things and beginning to follow them with his gaze, she will as a rule hit upon defective sight as the explanation; and at this stage an ophthalmologist will probably be sought.

Ophthalmoscopy

Whether it be a question of trying to differentiate between ocular and mental defect, or of obtaining some neuro-retinal clue to general diagnosis, or of allaying suspicion when the question of an eye lesion has been raised, I am strongly in favour of examining infants under general anaesthesia. The suggestion is not always well received, but there are powerful arguments in favour of it unless the baby is too ill—in which case we must do what we can without such aid, and aim at a general anaesthetic later on. During my earlier years in ophthalmology I wasted a lot of time trying to get precise information about babies' fundi without anaesthesia. After a quarter of an hour's patient concentration I often found the utmost I could honestly report was: "Nothing abnormal detected."

But this is not enough with the kind of patient that I chiefly have in mind. I am thinking of the situation that arises when the family doctor has reason to suspect eye trouble or mental defect, or both, to account for backwardness in development. In such circumstances ophthalmoscopy can often yield information without which it is impossible to build up a realistic prognosis. Unless we can make a thorough survey of the fundi we run the risk of missing signs which the parents ought to be told with the least possible delay. Therefore it is not enough to say: "Nothing abnormal detected." We should either describe what we find wrong, or else simply say: "Fundi normal."

In most cases the examination under general anaesthesia need not be repeated. If the fundi are established as normal, well and good. Many of the abnormal things detected are permanent, non-progressive, and not amenable to topical therapy; nevertheless it is important that they should be recorded, and taken into consideration when an infant's future is being planned. Before the second world war I used to make an occasional examination under general anaesthesia in the out-patient department, but that was never a satisfactory arrangement. It is not always easy to find an anaesthetist at short notice, and there are many obvious

objections to the ophthalmologist administering the anaesthetic, although I must admit that I have often functioned in the dual role. The greatest objection of all against out-patient general anaesthesia is the risk to the baby afterwards. I never met with any tragic outcome, but I believe there is a danger, even if it be slight, in sending a small baby out into the streets and crowded vehicles just after a general anaesthetic. Under in-patient conditions, however, the risks of a few minutes' anaesthesia are almost nil, unless life is already at a very low ebb, in which case the examination would of course be postponed. My own practice is to get the baby into hospital the day before the anaesthetic, and then he can go home the day after examination if no additional investigations are contemplated.

At the beginning of the examination under general anaesthesia we should inspect the anterior segment of each eye with the aid of a loupe, because fundus lesions are not uncommonly associated with other defects of the eye. Then we note the main features of the optic disk, follow the main vessels out to the periphery, and scrutinize the macular region. Many people like to insert a speculum to facilitate ophthalmoscopy, but I hardly ever do so nowadays, because the exposed cornea so soon desiccates. That necessitates either taking the speculum in and out at intervals, which is a nuisance, or repeatedly instilling drops of normal saline, which is not only a nuisance but also blurs the ophthalmoscopic picture. Without a speculum, I can easily hold the eyelids open with two fingers, and I keep the baby's cornea moist and lubricated by means of his own natural secretions. That is to say, my fingers draw the lids closed every few seconds. A colleague criticized my method on the ground that the baby's eyes are usually elevated under general anaesthesia. "The main advantage of a speculum is," he said, "that it enables you to apply your fixation forceps to draw the eye down." My retort was that a properly anaesthetized baby's eyes seldom shoot upwards, and I hardly ever resort to fixation forceps for this examination.

Interpretation of Ophthalmoscopic Signs

I have already referred to Tay-Sachs disease and toxoplasmosis as examples of diseases with ocular complications. The phakomatoses also form an interesting group in which eye defects may link up with intracranial lesions. Another fascinating subject is rubella, but I cannot dwell on all these things here.

My next task is to underline some of the commoner fallacies and mistakes associated with the ophthalmoscopic examination of babies.

1. Whenever I delve into my memories of other people's notes on infantile fundi, one impression stands out clearly; it is: *For every genuine abnormality that escapes detection, at least a dozen imaginary ones are described.* I wish I could more often meet with that simple comment "Fundi normal," but I am driven to conclude that many people are afraid to commit themselves so far, lest some other person should afterwards discover a lesion; or else they misinterpret something so as to make it fit in with what they vaguely remember from a textbook. Incidentally some observers evidently cannot believe that a blank stare is compatible with a pair of normal eyes; but surely there is nothing paradoxical about the association. In gross widespread injury or maldevelopment of the brain we should naturally expect the occipital lobes and various other regions concerned with vision to be implicated, so that there is no need for us to invent optic atrophy to explain the lack of sight in an infant thus afflicted.

2. *Optic disk pallor* is a potent source of confusion. It is widely recognized that the colour of healthy optic disks can vary within wide limits, but there are still many people who do not appreciate the extent of that variation, and many others who fail to realize that the average *infantile* disk is paler than that of a child or adult. Therefore many babies have been wrongly described as suffering from optic atrophy. It is in fact impossible to know the *degree* of a

partial optic atrophy until the subject is old enough to perform visual acuity and visual field tests.

3. *Blurred Disk Margins.*—Although it has been repeatedly pointed out that mere blurring does *not* mean swollen disks, this error keeps raising its head. Genuine swelling is always accompanied by corroborative signs, which I need not here describe in detail, but I should like to stress that many a perfectly healthy child is running about without any distinct margin to his optic disks.

4. *Congenital connective-tissue vestiges* on and near the optic disk are in most cases harmless, but they often create alarm and uncertainty among inexperienced users of the ophthalmoscope.

5. *Retinal hypoplasia* is normal in the early months of extrauterine life. Thus the pattern of the choroido-retinal vessels stands out distinctly in a larger proportion of babies than it does in children's or adult's fundi. Moreover, the macular stippling may be disquietingly conspicuous even in the absence of any degenerative change.

Correlation of Fundus Signs with the Other Medical Data

As the optic nerve is part of the central nervous system it is not surprising that optic atrophy or hypoplasia should so often be associated with low-grade mentality. Such major defects can arise from faulty genes, from intrauterine disease, or from birth injury, and we cannot always get conclusive evidence about the factors which have been responsible in one particular instance. From the standpoint of prognosis, however, these cases of blindness associated with gross mental defect present a relatively simple problem; and the doctor's main task is to console the parents and to give them advice about the baby's upbringing.

At the opposite extreme are various minor defects not likely to interfere with ordinary activities and not requiring any action other than a note in the medical records. Between these extremes a wide range of clinical problems still arise, demanding careful consideration by all concerned. Optic atrophy or gross macular degeneration affecting each eye obviously necessitates education by special methods, even if the patient is mentally normal and free from other physical defects, but in practice many borderline situations arise. I have already referred to the late starter who is suffering from slowly mending birth trauma to the brain, and who ultimately turns out far better than we could at one stage expect.

Another possibility to bear in mind is delayed myelination of the optic nerve, described by several French authors under the heading of "la pseudo-atrophie grise du nerf optique." Myelin is essential for the conduction of light-impulses, so that retarded myelination is in certain cases the reason why a baby apparently blind goes on to develop normal sight. Therefore no attempt should be made to give final or comprehensive advice about the disposal of infants as soon as any defect is apparent. It is, of course, important that some kind of prognosis, however provisional and incomplete it may be, should be given to the parents without delay. Most parents will appreciate how many uncertain factors must be weighed, and they will go on hoping for progress until the position clears.

It is impossible to lay down a set of rules governing the disposal of handicapped infants, whether the handicap be visual or mental or combined. From one point of view the mother is far and away the best person to look after a child with a disability, but that solution is not always feasible. The mother may lack the necessary qualities of patience and courage; or, on the other hand, she may tend to devote herself so utterly to a helpless infant that she has to neglect the rest of the household. The family doctor with all his knowledge of the home background is obviously better qualified than the ophthalmologist to give advice about disposal, but many problems can best be settled by joint discussion.

One of the questions often raised by the parents of a defective child is the advisability of having other children; and the answer depends mainly on the nature of the defect. Tay-Sachs disease, epiloia, and Schilder's disease are strongly familial, and in such conditions we can emphatically advise against any future pregnancy. Mongolism, on the other hand, is most unlikely to occur twice in the same marriage, although isolated instances are on record. In cases of primary amentia with no history of any similar trouble in a near relation the chances of another baby being affected are probably not more than 5%; but 5% is a lot when one considers the appalling consequences, and most couples would hesitate to take the risk. Now that so many statistics have been carefully worked out by experts in the field of heredity, the advice of a geneticist will often be helpful in answering queries about further children.

Concerning the relationship between fundus lesions and mental capacity, I want to make one final point, and that relates to a baby's future visual acuity. We must avoid giving too gloomy a forecast about the sight unless fundus lesions are very severe, because it is astonishing how resourcefully some children learn to interpret blurred images if their mind is bright.

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PRESENTATION TO LORD INMAN SERVICES TO CHARING CROSS HOSPITAL

To mark his 36 years' work for Charing Cross Hospital and Medical School, Lord INMAN was presented with his portrait at a ceremony at the Royal College of Surgeons on Tuesday. Lord Inman is chairman of the hospital. The presentation was made by Lord Justice MORRIS, a fellow member of the board of governors, and Mr. JOHN ADAMSON, the deputy chairman of the board, presided. The portrait, which will eventually hang in the board room at Charing Cross Hospital, is by Mr. Norman Hepple, A.R.A., and a replica of it was given to Lady INMAN.

A Notable Administrator

In 1921, at the early age of 29, Lord Inman, then Mr. Philip Inman, was appointed house governor of Charing Cross Hospital from 583 applicants. At the time he was chief officer of the Hospital Saturday Fund.

In the days of the voluntary hospitals the chief administrative officer had a dual function. Not only had he to manage the hospital but he had also to obtain the income necessary for its existence. Charing Cross was one of the relatively few hospitals able to balance its accounts, thanks

to its house governor's great success in raising funds. From the time of his appointment until the inception of the National Health Service, Lord Inman was instrumental in raising £1.5m. for the hospital.

When in 1936 the then chairman of the hospital died, the hospital council invited Mr. Inman to succeed him. Shortly after his appointment as chairman of the hospital,