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# An international classification of retinopathy of prematurity

PREPARED BY AN INTERNATIONAL COMMITTEE*

## I. Introduction

As a result of advances in technology, particularly in life support systems capable of keeping tiny premature infants alive, and better observation of the premature infant fundus with improved ophthalmoscopic techniques, including the indirect ophthalmoscope, much has been learned about the early active stages of retinopathy of prematurity (ROP). This term is preferred because it can be utilised to describe all phases of the retinal changes observed in premature infants. The traditional term, retrolental fibroplasia, is inappropriate in the acute phase of this disorder, for it describes solely those later cicatricial changes which involve the eyes of only the most severely affected infants. Much of what has been learned over the past two decades about the disease in its modern form fails to fit with the Reese classification system[1] or any other classification system extant. Furthermore, the real incidence of the disease may be increasing, though the evidence on this point is inconclusive, and treatment of the disease in its active and cicatricial form has been advocated, but it is not always clear what disease stage is being treated and what the results of such treatment are. Hence the need for a new classification system of the acute stages of ROP at this time, with a classification of the cicatricial stages to follow.

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## II. The classification

The system presented here differs from previous systems in that it permits the examiner to specify at the outset two parameters of the disease not recognised in other classification systems. These are the *location* of the disease in the retina and the *extent* of the developing vasculature involved. In addition the examiner grades the retinopathy according to a system more consistent with current clinical observations.

### A. LOCATION

For the purpose of defining this variable, three zones of retinal involvement are recognised (Fig. 1). Each

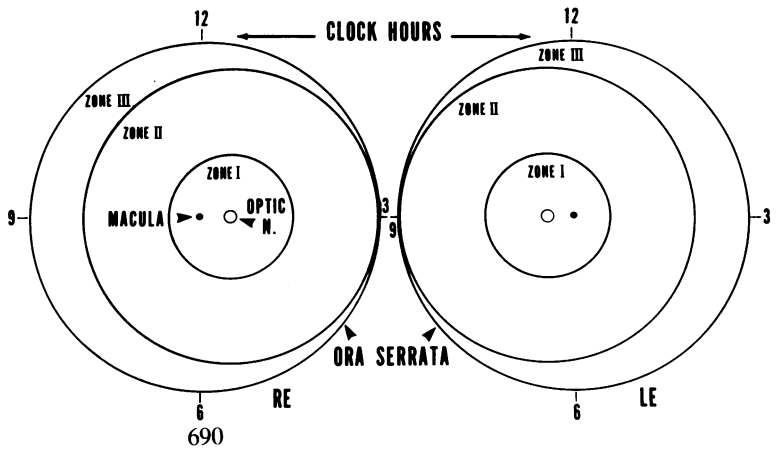



Fig. 1 *Schematic illustration of the retina of the right and left eye showing the zone borders and the clock hours employed to describe the location and extent of ROP.*

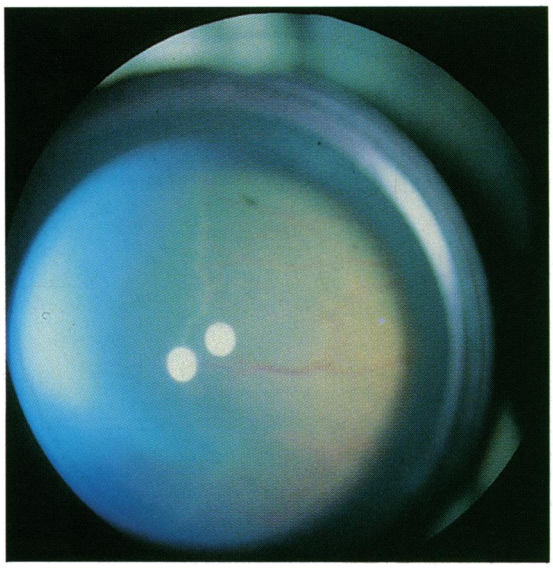


Fig. 2

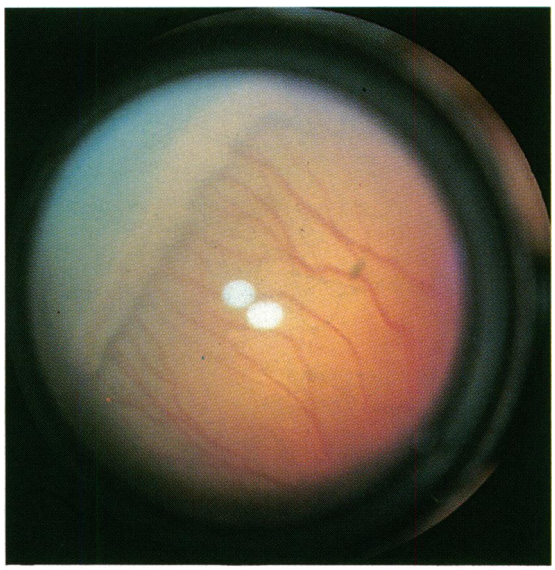


Fig. 4

zone is centred on the optic disc rather than the macula, contrary to standard retinal drawings. The new scheme was selected because normal retinal vascular growth proceeds outward from the disc toward the ora serrata in an orderly fashion. The first two zones occupy that portion of the fundus which lies behind a circle drawn using the disc as the centre and the distance to the nasal ora serrata at the horizontal meridian as its radius. Therefore any ROP which is circumferential must by definition fall into one of these two posterior zones.

Zone I (posterior pole or inner zone) consists of a circle (Fig. 1) the radius of which subtends an angle of 30° and extends from the disc to twice the distance from the disc to the centre of the macula. The limits of the zone are consequently defined as twice the disc-fovea distance in all directions from the optic disc, i.e. an arc of 60°.

Zone II extends from the edge of zone I peripherally to a point tangential to the nasal ora serrata (at 3 o'clock in the right eye, 9 o'clock in the left) and round to an area near the temporal anatomic equator.

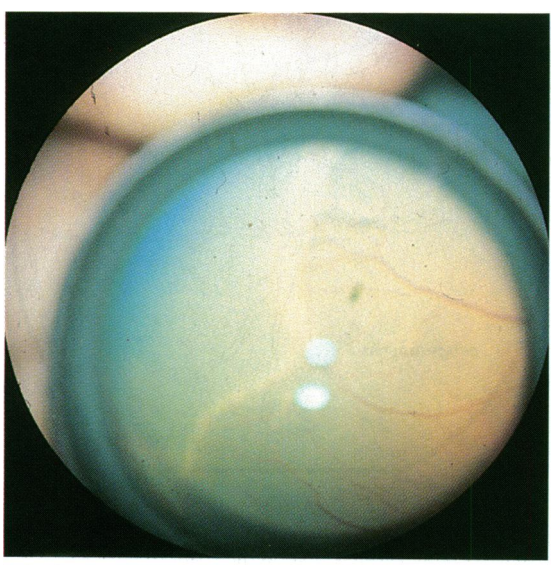


Fig. 3

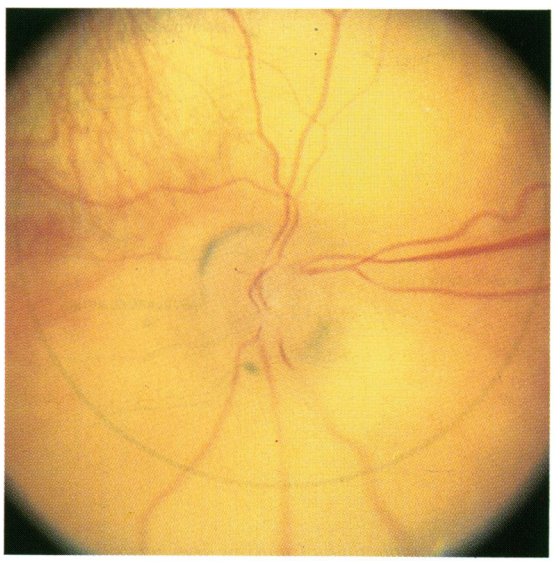


Fig. 5

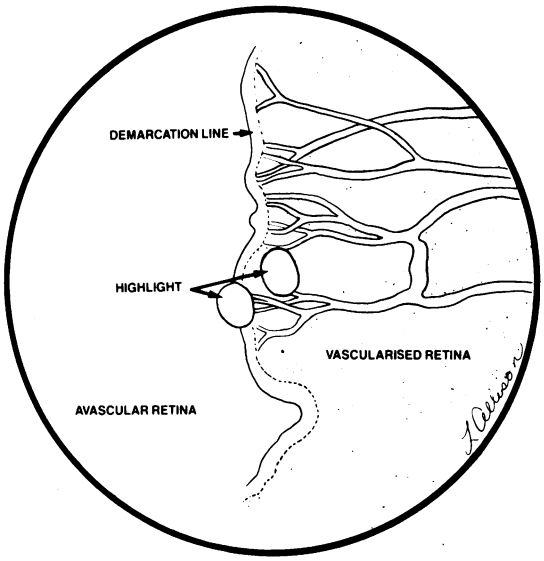



Fig. 2

Fig. 2 *Fundus photograph and line drawing to illustrate the demarcation lines of stage 1.*

The temporal edge of zone II cannot be more accurately defined clinically, as the anatomical landmarks needed to identify the equator in a premature infant are obscured. Indeed, these landmarks are sufficiently varied in humans to render precise locations difficult at any age.

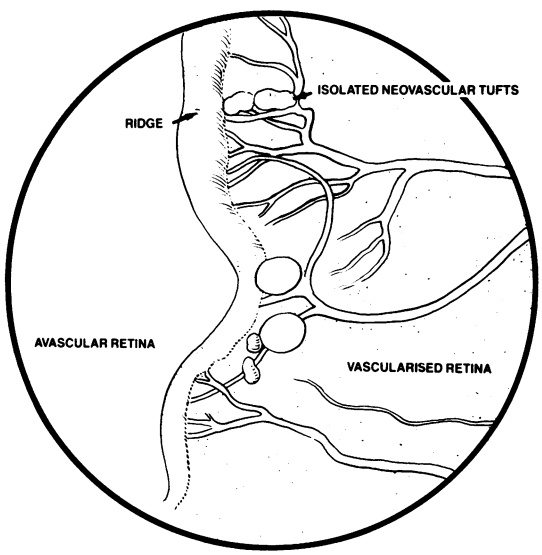



Fig. 3

Fig. 3 *Fundus photograph and line drawing to illustrate the development of the ridge characteristic of stage 2.*

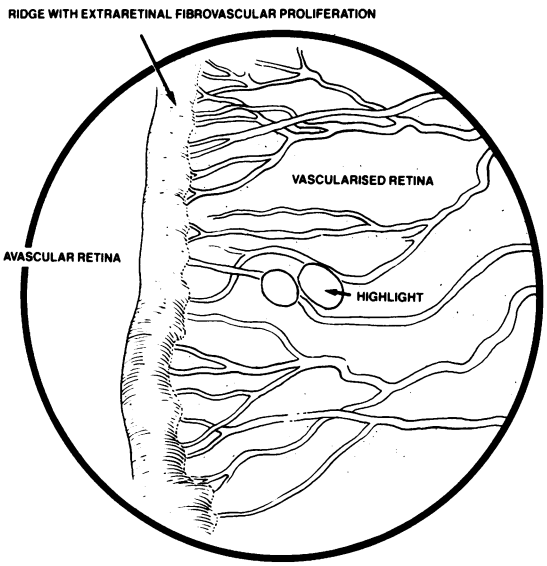



Fig. 4

Fig. 4 *Fundus photograph and line drawing of the extraretinal fibrovascular proliferative tissue of stage 3.*

Zone III is the residual crescent of retina anterior to zone II. This is the zone that is last vascularised in the premature eye and it is the zone, by common agreement of clinicians, most frequently involved with ROP.

B. EXTENT OF THE DISEASE (Fig. 1)

This is specified as hours of the clock. As the observer

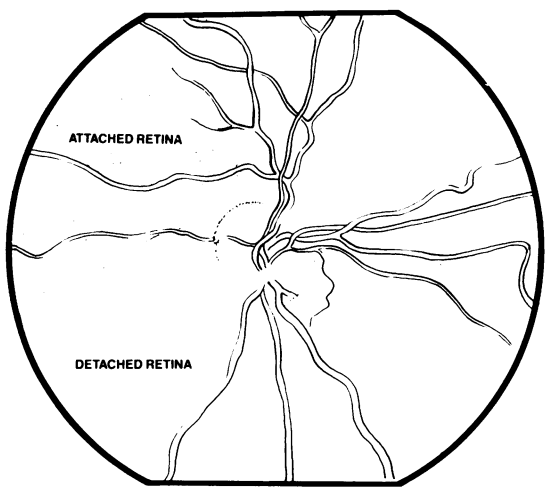



Fig. 5

Fig. 5 *Fundus photograph and line drawing of the shallow exudative retinal detachment characteristic of stage 4 involvement.*

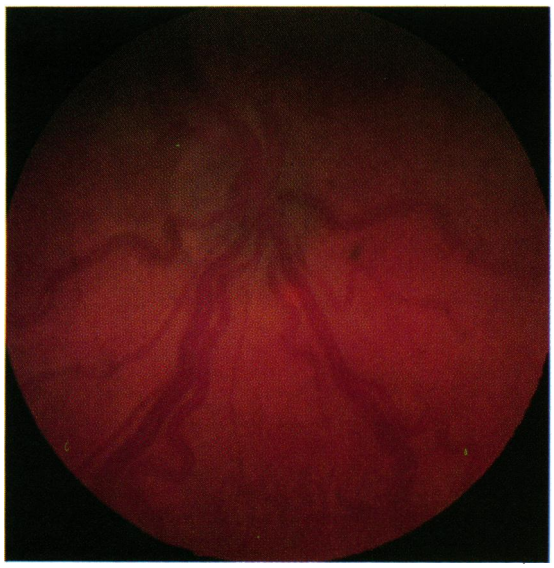


Fig. 6 *Fundus photograph of the posterior venous dilatation and arteriolar tortuosity characteristic of 'plus' disease.*

looks at each eye, 3 o'clock is to the right and nasal in the right eye and temporal in the left eye, and 9 o'clock is to the left and temporal in the right eye and nasal in the left eye.

### C. STAGING THE DISEASE

In addition to the above two parameters the final one to be specified is the level of abnormal vascular response observed. Here four stages are recognised,

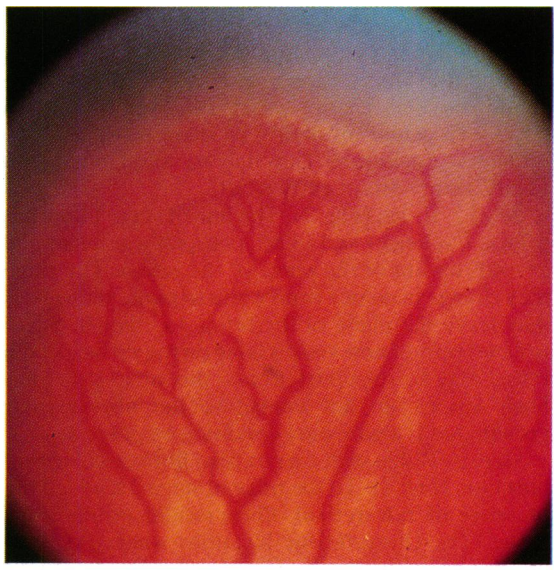


Fig. 9

and staging for the eye as a whole is by the most severe manifestation present. However, for purposes of recording the complete examination each stage is defined and the extent of each stage by clock hours is recorded.

*Stage 1.* Demarcation line (Fig. 2). This line is a thin but definite structure that separates the avascular retina anteriorly from the vascularised retina posteriorly. There are recognisable abnormal branching or arcading of vessels leading up to it. It is relatively flat, lies within the plane of the retina, and

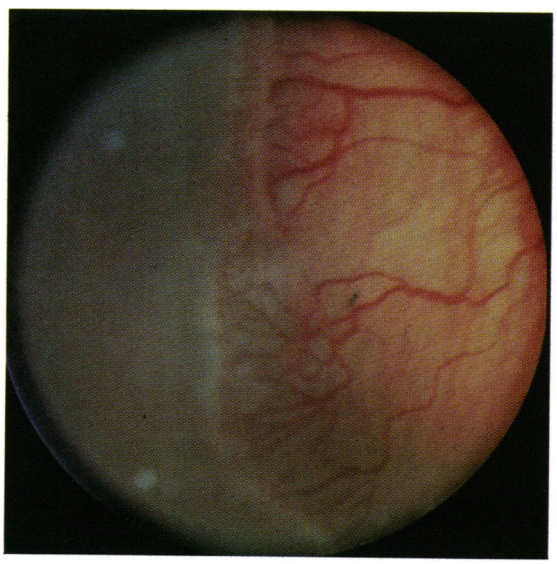


Fig. 8

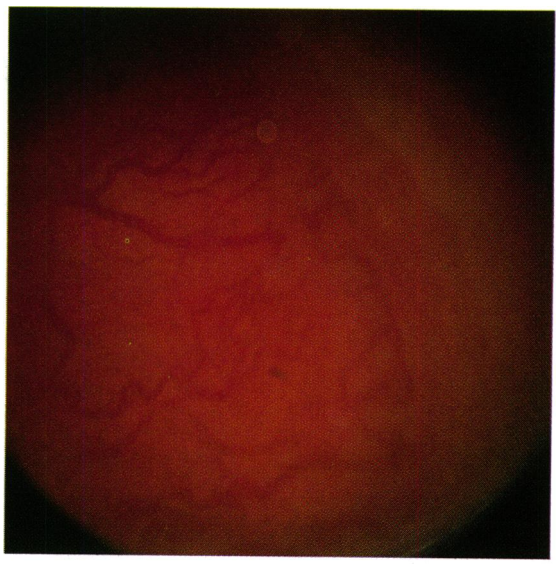


Fig. 10

is white in color. There have been described vascular changes that can be apparent prior to the development of the demarcation line. However, these more subtle vascular changes vary considerably, cause no known ocular morbidity by themselves, and are difficult to quantitate. They may be noted, but do not justify a diagnosis of early ROP.

*Stage 2.* Ridge (Fig. 3). The line of stage 1 now has grown, has height and width, occupies a volume, and extends up out of the plane of the retina. The ridge may change in colour from white to pink, and vessels may leave the plane of the retina to enter it. Small isolated tufts of new vessels lying on the surface of the retina may be seen posterior to this ridge structure. Such lesions do not constitute the fibrovascular growth that is a necessary condition for stage 3.

*Stage 3.* Ridge with extraretinal fibrovascular proliferation (Fig. 4). To the ridge of stage 2 is added the presence of extraretinal, fibrovascular proliferative tissue. The characteristic locations where this proliferating tissue may be found are: (1) continuous with the posterior aspect of the ridge, causing a ragged appearance of the ridge as proliferation becomes more extensive; (2) immediately posterior to the ridge but not always appearing to be connected with it; (3) into the vitreous perpendicular to the retinal plane. Fibrovascular proliferation may be seen in any or all of these locations in stage 3 ROP.

*Stage 4.* Retinal detachment (Fig. 5). To the above is added unequivocal detachment of the retina. It may be caused by an exudative effusion of fluid, traction, or both, even in this early stage. In any case, the examiner should specify its location, extent, and nature. It may be particularly difficult to differentiate shallow posterior retinal detachments, as the loss of choroidal pattern may be subtle and difficult to distinguish through the increasing vitreous haze of severe disease. Serial examinations may be required to be certain of a true detachment. It should be emphasised that the presence of elevated retinal vessels running from the retinal plane to the height of the ridge does not constitute a posterior detachment.

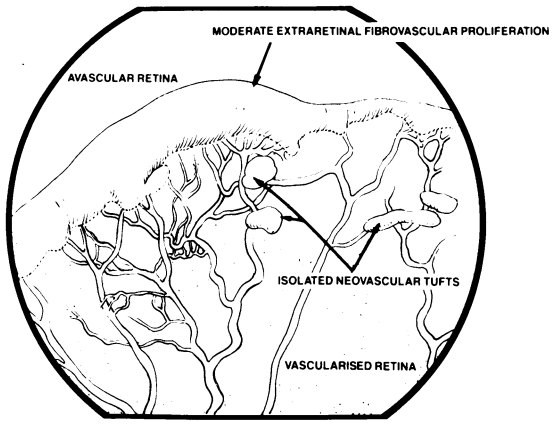



Fig. 9

Fig. 9 *Fundus photograph and line drawing of 'moderate' proliferation of extraretinal fibrovascular tissue from the ridge.*

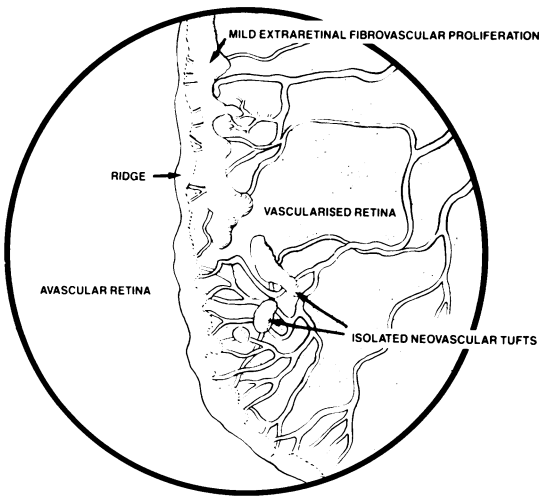



Fig. 8

Fig. 8 *Fundus photograph and line drawing to illustrate the amount of extraretinal fibrovascular proliferative tissue judged to be 'mild' stage 3.*

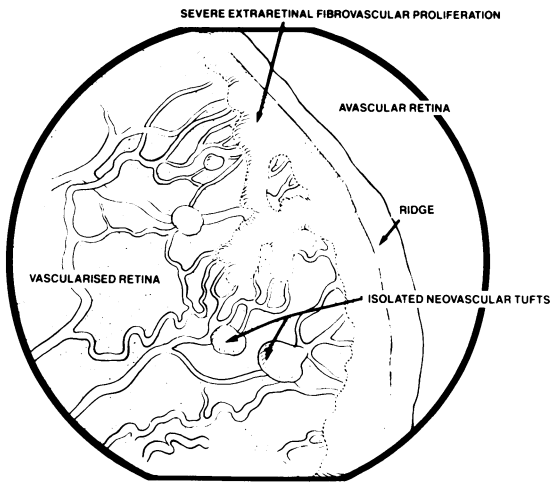



Fig. 10

Fig. 10 *Fundus photograph and line drawing of extraretinal fibrovascular proliferation of amounts of tissue judged to be characteristic of 'severe' stage 3 ROP.*

RETINOPATHY OF PREMATURITY (ROP) OPHTHALMIC EXAMINATION RECORD

BIOGRAPHICAL DATA

Name_____ Hospital # ___ ___ ___ ___ ___ ___ ___

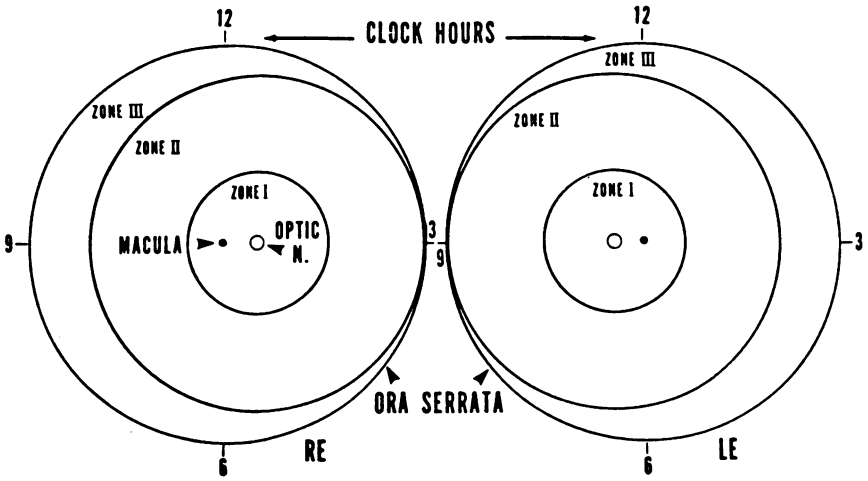
Birthdate (MM/DD/YY) ___ ___/___ ___/___ ___ Sex (M=1, F=2) ___

Birthweight (grams) ___ ___ ___ ___ Gestational Age (weeks) ___ ___

Multiple Births (Single=1, Twin=2, Triplet=3) ___

EXAMINATION

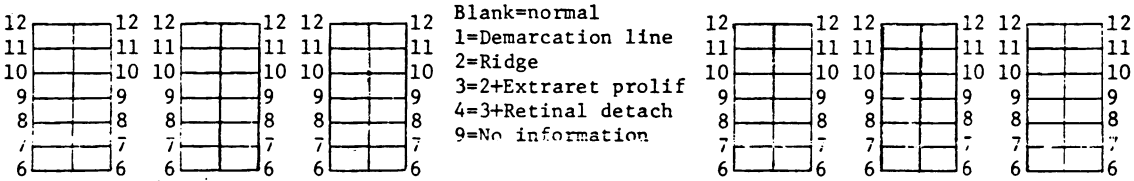
Date of Exam ___ ___/___ ___/___ ___ Examiner's Initials or # ___ ___ ___




ZONE
Mark with 'X'

Z 1 ☐ Z 2 ☐ Z 3 ☐ Z 1 ☐ Z 2 ☐ Z 3 ☐

STAGE AT CLOCK HOURS




Blank=normal
1=Demarcation line
2=Ridge
3=2+Extraret prolif
4=3+Retinal detach
9=No information

Mark highest stage at every clock hour

☐ If Stage 3: 1=mild, 2=moderate, 3=severe ☐

☐ If Stage 4: 1=exudative, 2=tractional, 3= combined ☐

Other Findings
Mark with 'X'

O.D. O.S.

☐ A Dilatation/tortuosity posterior vessels ☐

☐ B Iris vessel dilatation ☐

☐ C Pupil rigidity ☐

☐ D Vitreous haze ☐

☐ E Hemorrhages ☐

Cicatricial RLF (Reese, 1953)

Mark with 'X'

O.D. O.S.

☐ I. Small mass opaque tissue in periphery without detachment ☐

☐ II. Larger mass opaque tissue in periphery with localized detachment ☐

☐ III. Larger mass in periphery with traction fold to disc ☐

☐ IV. Retrolental tissue covering part of pupil ☐

☐ V. Retrolental tissue covering entire pupillary area ☐

COMMENTS:

Signature

Fig. 7 *The suggested format of the ophthalmological examination record to permit complete recording of the detailed examination results both graphically employing the retinal drawing and numerically for later analysis if desired.*

### III. 'Plus' disease

Progressive vascular incompetence, occurring along with the changes described at the edge of the abnormally developing retinal vasculature, is noted by increasing dilatation and tortuosity of the peripheral retinal vessels, iris vascular engorgement, pupillary rigidity, and vitreous haze. When, and only when, the vascular changes are so marked that the posterior veins are enlarged and the arterioles tortuous, then the designation 'plus' is added to the ROP stage number (Fig. 6). For example, the ridge of stage 2 ROP combined with posterior vascular dilatation and tortuosity would be written, stage 2+ ROP. When the ROP is located in zone I or posterior zone II and 'plus' disease is present, progression may be very rapid.

### IV. Recording the results

For purposes of recording the results of the examination, the appended examination record is recommended (Fig. 7). The scheme is computer compatible.

### V. Problems confronted

The committee recognises that no classification, including the present one, is perfect. During the course of our deliberations several problem areas were encountered for which approximate solutions were developed realising that, with time and experience in the use of the classification, better solutions for its users will emerge. The problems were:

*(a) Definition of zone.* Anatomical landmarks other than the disc and the ora may be difficult to discern in the premature eye, and therefore the boundaries of the zones, I and II for example, are only approximate. The same can be said of zone II and III, except that, if vascularisation has reached the nasal ora, any disease found elsewhere is by definition in zone III. The committee recommends that, where doubt exists as to the appropriate zone to locate the disease, it be located in the more posterior zone.

*(b) Stage 3 disease.* The committee clearly recognises the need to further subdivide stage 3 disease for its potential prognostic significance. To do so it chose as its yardstick the amount of fibrovascular proliferative tissue present. If only limited amounts can be recognised by the examiner, this would constitute 'mild' stage 3 (Fig. 8). If on the other hand significant amounts of tissue are seen infiltrating the

vitreous, proliferating posterior from the ridge, then this is 'moderate' stage 3 (Fig. 9). Finally, if massive infiltration of the tissues surrounding the ridge is occurring, the threshold for 'severe' stage 3 has been reached (Fig. 10).

*(c) Overlap with cicatricial disease.* It is clearly recognised that in this classification traction detachment forms part of the description of stage 4. Classically this has been reserved for the cicatricial phase of the disease. Retinopathy of prematurity is a continuum and not easily fitted into any arbitrary man-made scheme. Nevertheless, the description of stage 4 would be incomplete without allowing for the occurrence of traction detachments as part of it. For the time being the committee recommends the retention and use of the Reese classification of cicatricial disease[1] to describe disease changes beyond those described in this classification.

## VI. Sponsorship

The classification is the product of the joint effort of 23 ophthalmologists from 11 countries. Though the committee was an ad-hoc body, it obtained sponsorship for its deliberations from the American Academy of Ophthalmology, the American Academy of Pediatrics, the American Association of Pediatric Ophthalmology and Strabismus, the National Eye Institute, the Division of Maternal and Child Health of the Bureau of Health Care Delivery and Assistance, the March of Dimes, the Alberta Heritage Foundation for Medical Research, and Ross Laboratories. In no small measure this support has provided the encouragement necessary to complete work on this classification. The success or failure of this classification will be judged by its use within the ophthalmological and paediatric communities.

## VII. Summary conclusions

The unifying principle underlying this classification system is: the more posterior the disease and the greater the amount of involved retinal vascular tissue, the more serious the disease. The staging of the disease at any given location expresses the natural history and evolution of events at the border between vascularised and avascular retina. The classification system is designed to permit the examiner full latitude in transcribing his observations so that they will be immediately intelligible to another examiner who may not have had the opportunity to examine the specific infant.

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### Reference

1 Reese AB, King MJ, Owens WC. A classification of retrolental fibroplasia. *Am J Ophthalmol* 1953; **36:** 1333–5.