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The Recognition of Active Retrolental Fibroplasia

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As the incidence of retrolental fibroplasia increases, physicians must become familiar with its appearance. This report describes the examination techniques and morphology of the active and regressive stages of retrolental fibroplasia.

After retrolental fibroplasia (RLF) was described by Terry in 1942 (1), it rapidly became the single largest cause of infant blindness in the United States. A national cooperative study (2) designed to seek causes demonstrated that the length of time the premature infant is kept in an oxygen-enriched environment is an important factor in causing RLF. As a result, the use of oxygen for premature infants was severely curtailed for several years.

In the ensuing years, however, pediatricians tended to use oxygen more liberally to manage idiopathic respiratory distress syndrome, and the incidence of RLF increased (3). Consequently, ophthalmologists are now often called upon to screen premature infants.

Infant Screening

The infant at greatest risk of developing RLF is one with a birth weight of less than 1500 gms (3.3 lb) who suffers from the respiratory distress syndrome with multiple episodes of apnea (3). All infants with a birth weight of less than 2000 gms (4.2 lb) or a gestational age under 34 weeks should be screened if they received oxygen in concentrations greater than room air.

Because ophthalmoscopic monitoring of vasospasm is inaccurate and early examination is impeded by vitreous haze and embryologic vascular remnants (Fig. 1), the best time to perform the examination is just before the infant is discharged from the nursery.

Examination Techniques

The ocular fundus cannot be examined properly unless the pupil is dilated. Tropicamide (½%) and phenylephrine (2.5%) are instilled in the conjunctival cul-de-sac 30 minutes and again 15 minutes before the examination takes place. Tropicamide relaxes the iris sphincter and phenylephrine stimulates the dilator muscle to produce maximal dilatation. Systemic absorption of tropicamide may produce lethargy or apnea. Phenylephrine may make the eyelid skin blanch, but it causes no adverse effects.

The binocular indirect ophthalmoscope is indispensable for viewing the retinal periphery. A wider view is obtained if a 2X condensing lens with a diameter less than 2.5 cm is used. Examination with this instrument may be combined with scleral depression to evaluate the retina as far anterior as the ora serrata.

The child may be examined within the Isolette if its body temperature is difficult to maintain (Fig. 2). Otherwise, the examination should be performed outside for better visualization. The infant will be more docile if examined soon after feeding. Its eyelids may be held apart with the fingers or a Sauer infant eyelid speculum (Fig. 3). If the speculum is used, the child's cornea must be kept moist with artificial tears.

If active disease is found, photographic documentation is helpful. Fluorescein angiography is a valuable research tool but is not used routinely. Both fundus photography and

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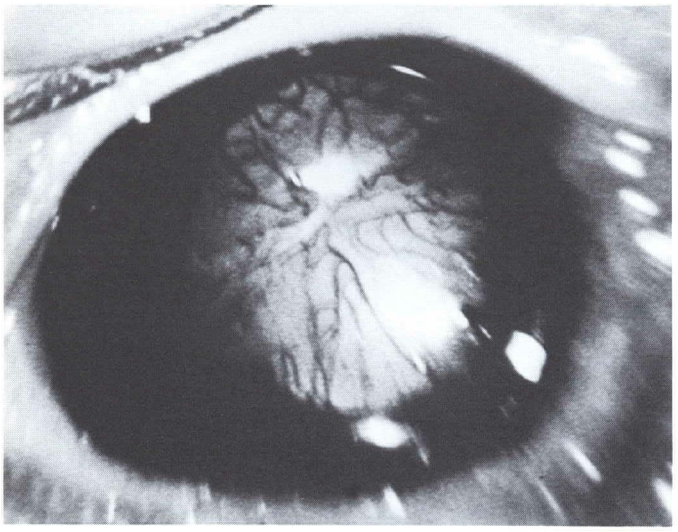

Fig. 1
Persistent Tunica Vasculosa Lentis.

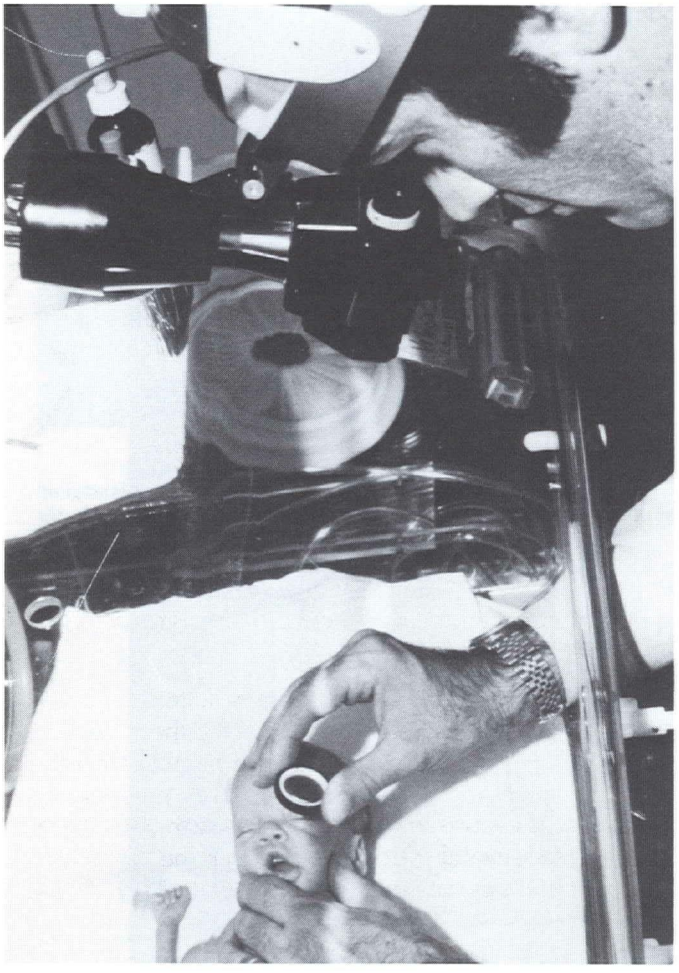

Fig. 2
Examination through clear dome of Isolette.

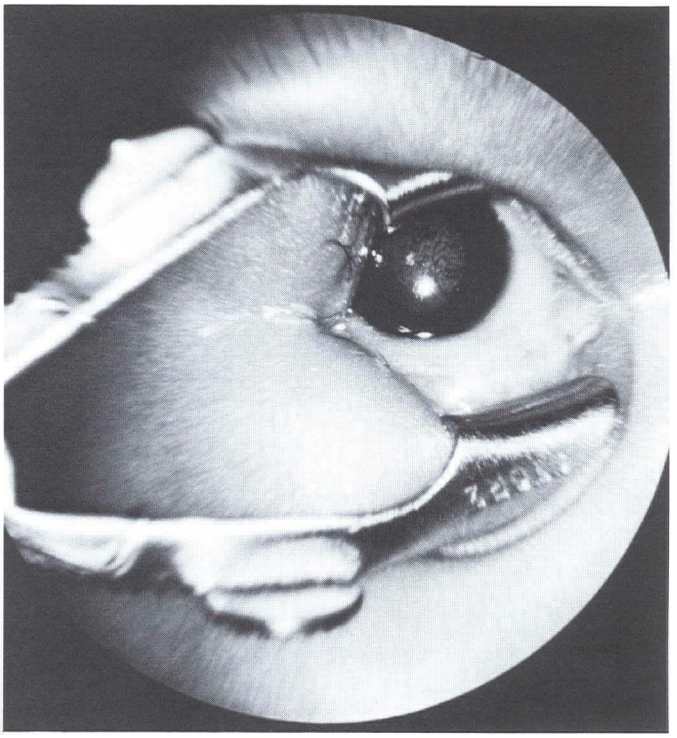

Fig. 3
Eyelids retracted with infant speculum.

fluorescein angiography are made easier by the pediatric photography platform designed to be used with the Zeiss fundus camera (4) (Fig. 4).

Recognition of Active Disease

The earliest definite clinical sign of RLF is tortuosity and dilatation of the vessels near the disc (Fig. 5), caused by increased blood flow to the abnormal vascular channels in the peripheral retina. The arteriovenous shunt, which develops at the junction of vascularized and avascular retina, is the most prominent structure in the fundus of the eye affected by RLF and is an important landmark to identify (Fig. 6). Neovascularization occurs near this structure, and normal vascularization begins here during the regressive stages. In full-term infants, the temporal periphery of the retina is normally not vascularized until after birth, so it is not surprising that in premature infants a wide avascular zone is present temporally. However, avascularity alone does not imply the presence of active RLF.

The major criterion for the diagnosis is a prominent demarcation line betwen vascular and avascular retina, with evidence of abnormal vascularization at that junction. Preretinal proliferative neovascularization develops immediately posterior to the arteriovenous shunt and is usually associated with areas of capillary nonprofusion (Fig. 7). It

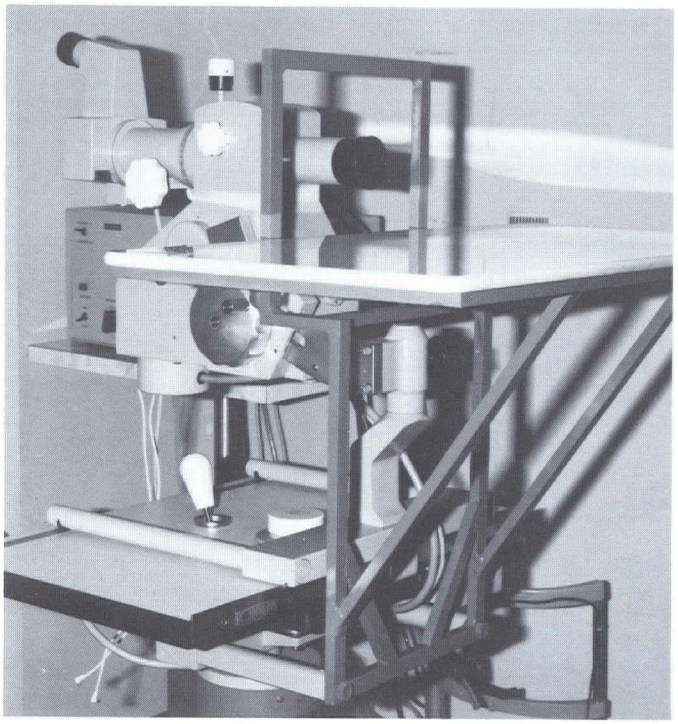

Fig. 4
Pediatric photography platform attached to Zeiss fundus camera.

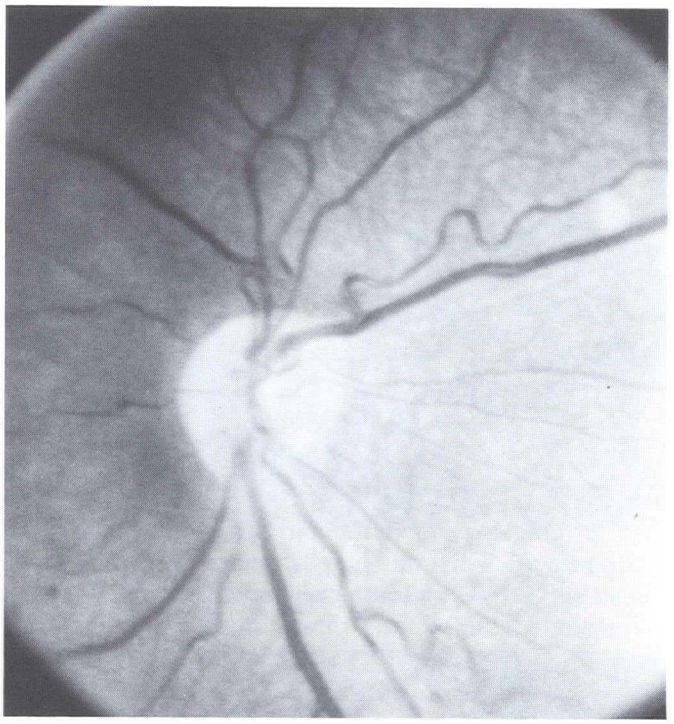

Fig. 5
Posterior pole of eye with active retrolental fibroplasia.

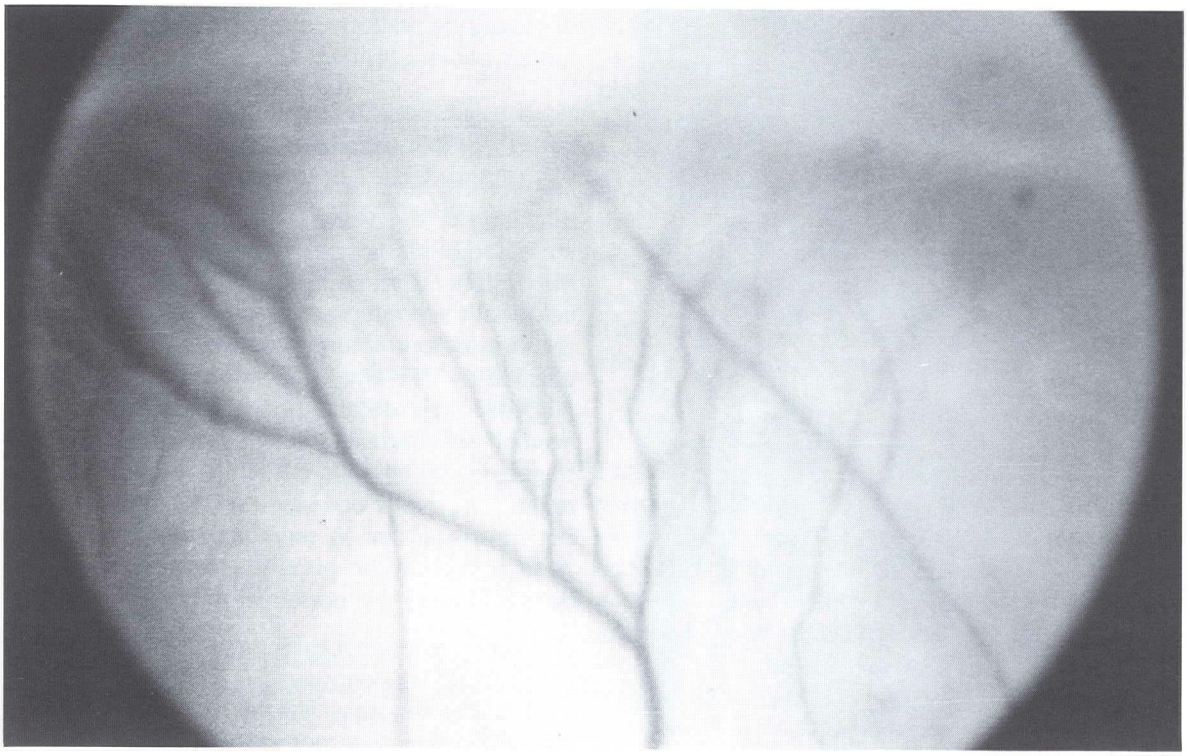

Fig. 6
Prominent arteriovenous shunt in temporal periphery with inserting retinal vessels.

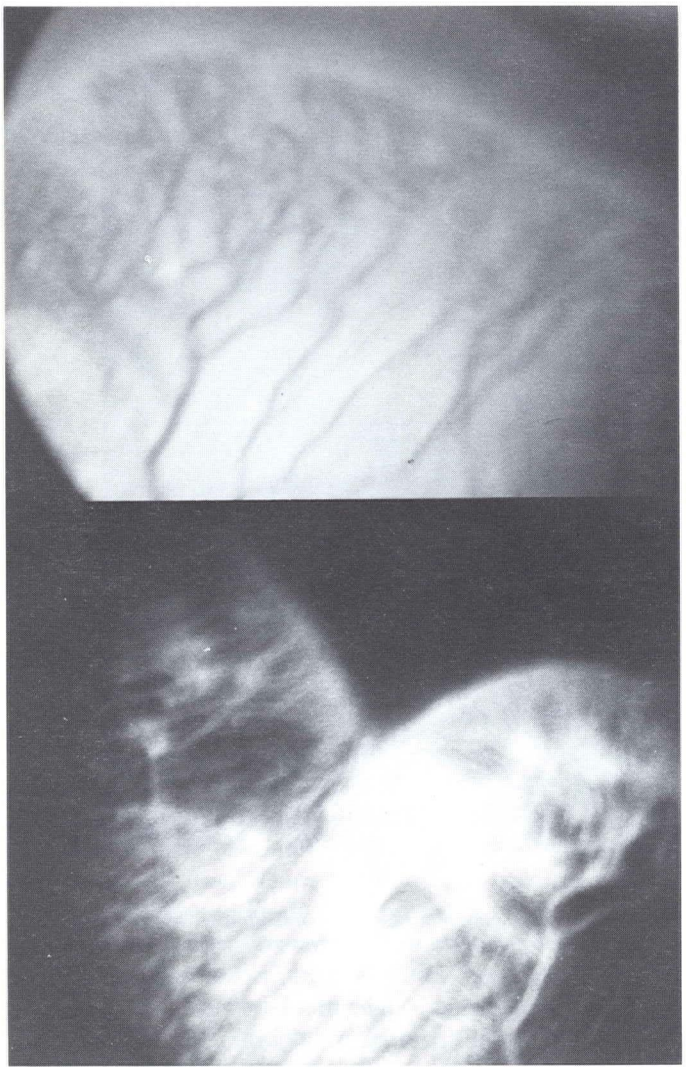

Fig. 7
Preretinal proliferative neovascularization (top); fluorescein angiography shows avascular retina, neovascularization, and capillary nonperfusion (bottom).

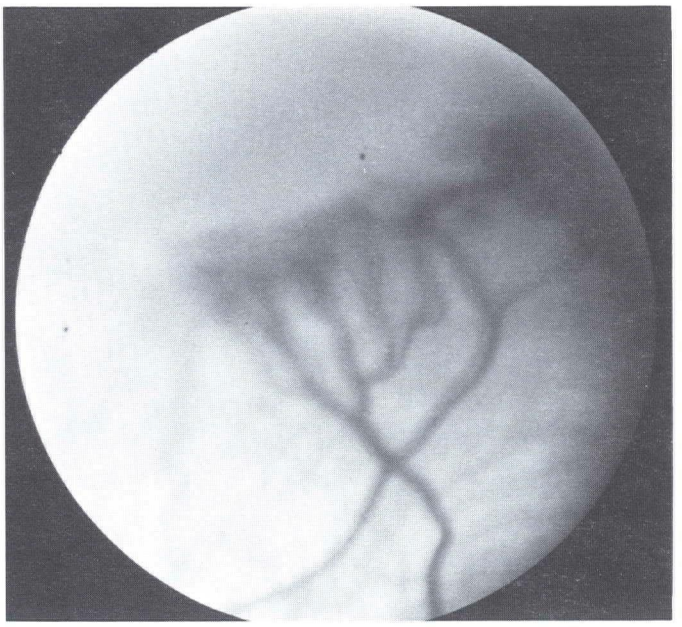

Fig. 8
Enlarged neovascular frond with hemorrhage along arteriovenous shunt.

looks like other peripheral proliferative retinopathies, especially those associated with the sickling hemoglobinopathies.

As the active phase develops into a more advanced stage, the neovascular fronds enlarge, hemorrhage appears near the fronds, in the vitreous and along the arteriovenous shunt, and the avascular retina may be thickened and elevated (Fig. 8).

It is important to recognize the early signs of regression of the active disease. O'Grady (5) first suggested that the beginning of the regression process may be signaled by a double demarcation line composed of the original arteriovenous shunt and a fine brush border of normal retinal vasculature. As vascularization becomes normal, the shunt is left behind and atrophies. Remnants may remain as a sentinel of the disease process (Fig. 9).

The role of the ophthalmologist is to advise physicians and parents of premature infants. Knowledge of the natural history of the disease process and recognition of its stages are paramount to accomplishing that goal.

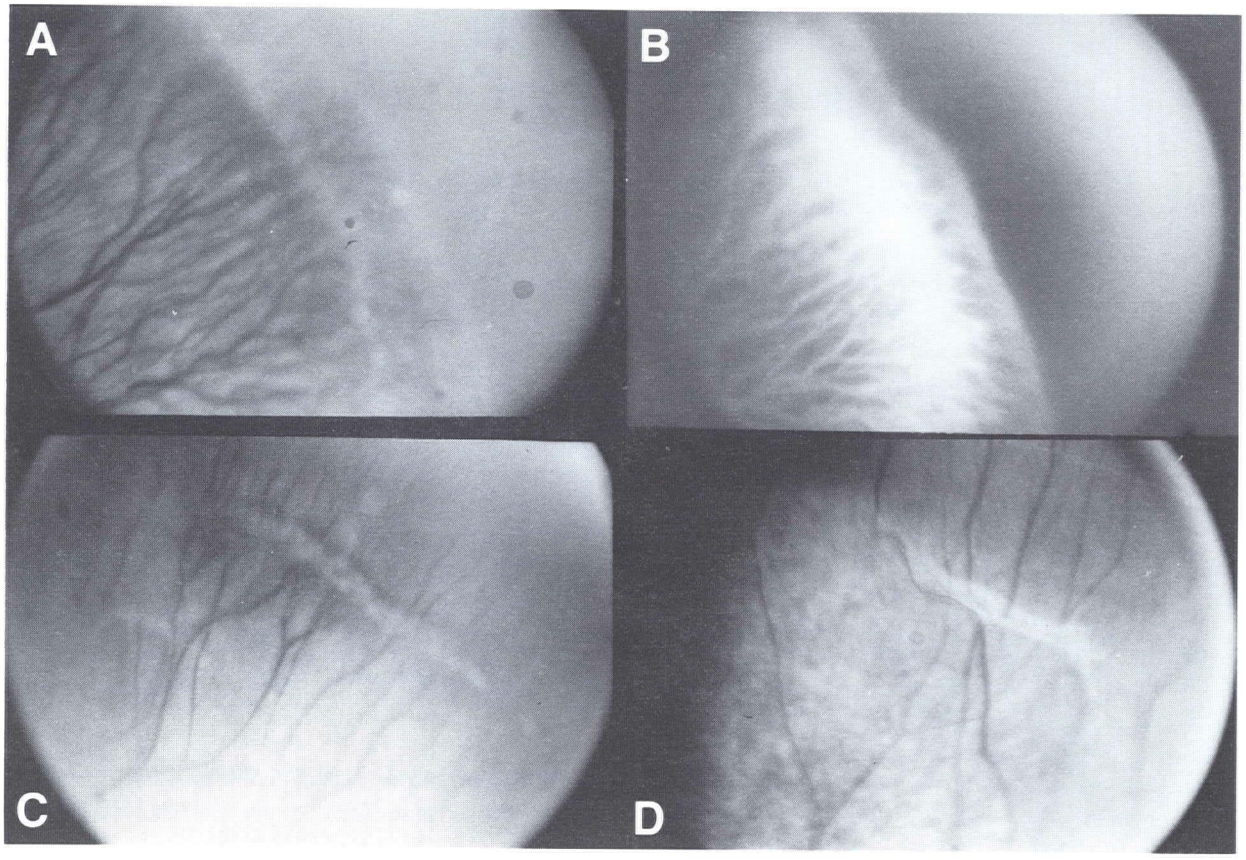

Fig. 9

A. Temporal periphery at age 10 weeks shows double demarcation line; B. Fluorescein angiogram shows fine brush border of normal retinal vessels; C. At 13 weeks, vessel growth has continued and shunt atrophy begins; D. At 16 weeks, only a gray remnant of the arteriovenous shunt remains.

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