In 1942 Terry described several children who had leukocoria and a clear lens. He postulated that this entity represented an overgrowth of the vascular tunica. Since then we have learned that this retinopathy occurs to a great degree in premature children. A plethora of factors have been incriminated over time, such as oxygen, sepsis, shifting of the oxygen curve, and bronchodyplasia. These children seem to be born with a fair amount of undeveloped retina, as vascularization is not completed until 1 month after birth. It is a switch from orderly angiogenesis to disorderly proliferation that produces the retinopathy of prematurity (ROP).

A classification based on the extent and position of the abnormality has been established. The position of the disease is denoted by three zones.

Zone 1: Distance from the optic disc to twice the disc macular distance in all directions
Zone 2: from the outer border of zone 1 to the nasal ora and to the equator temporally
Zone 3: from the outer border of zone 2 to the remaining ora

The extent of the disease is expressed as the hours of a clock. Staging denotes the amount of disease present.

- Stage 1: demarcation line
- Stage 2: ridge formation
- Stage 3: ridge with extraretinal proliferation
- Stage 4A: subtotal detachment—uninvolved macula
- Stage 4B: subtotal detachment—involved macula
- Stage 5: total retinal detachment

Using this classification, communications among investigators and treatment results are easier to comprehend. Cryotherapy has been suggested by the Cryotherapy Study Group. The threshold indications are five or more contiguous clock hours or 8 cumulative clock hours of stage 3 disease in eye zone 1 or 2 with plus disease. In patients with a retinal detachment encroaching on the macula, a scleral buckle should be considered. Children with stage 5 disease, may benefit from closed vitrectomy or open sky vitrectomy.
After the active disease has abated, cicatricial disease becomes evident. The vascular system is pulled temporally, with temporal pigmentation appearing later. These patients have perivascular and radially oriented lattice lesions, peripheral schisis, and break formation.

Fluorescein angiography in ROP patients typically demonstrates vascular leakage due to the extensive neovascularization. The electroretinogram changes as the retina detaches, with loss of the a-wave and then the b-wave. Visual field defects are variable in cicatricial disease depending on how much retina has been distorted.

Histologically, it has been theorized that the spindle cells (vascular precursors) undergo transformation from a vasoformative role to that of vasoproliferation. It is manifested by an increase in gap junctions between these spindle cells. In stage I disease the demarcation line is set up by arteriovenous collaterals. Multiple microvascular abnormalities can be seen in this region. As retinitis proliferans develops, the vessels grow through the internal limiting membrane into the subvitreal space and eventually into the vitreous. Contracture of these vessels can cause the hemorrhages, and the fibrocellular components parallel to the shunt cause contraction and elevation of the shunt.

Selected Reading


