The rationale for cryotherapy with a prophylactic scleral buckle for Zone I threshold retinopathy of prematurity

HELEN A. MINTZ-HITTNER & FRANK L. KRETZER
Cullen Eye Institute, Baylor College of Medicine, Houston, TX 77030, USA

Received and accepted 29 November 1989

Key words: cryotherapy, myofibroblasts (tractional forces), ocular growth, scleral buckling, spindle cells (angiogenic stimulation), Zone I retinopathy of prematurity

Abstract. Our current surgical protocol for Zone I threshold retinopathy of prematurity (ROP) has evolved over 15 years and is rationalized by increasing knowledge of two pathologic processes of ROP: 1) angiogenic stimulation of spindle cells (clinically invisible) near the vitreal surface of the avascular retina; and 2) tractional forces of myofibroblasts [clinically visible as extraretinal fibrovascular proliferation (EFP)] in the vitreous overlying the vascular retina. These two pathologic processes occur concomitantly with normal anterior ocular growth with a constant optic disc-macular distance. Our current surgical protocol for Zone I threshold ROP involves complex surgeries to achieve success defined as a macula which always remains anatomically attached, but which may be distorted or ectopic. This protocol requires cryotherapy in at least two sessions. The first is to the avascular retina to destroy spindle cells. The second is to the EFP to destroy myofibroblasts and to the shunt to eliminate the site of origin of myofibroblasts. The protocol also requires the concomitant placement of a prophylactic scleral buckle to allow formation of a new complete ora serrata while remnant myofibroblasts contract and while anterior ocular growth continues.

Our current protocol of surgical intervention for Zone I threshold retinopathy of prematurity (ROP) as defined by the USA multicenter CRYO-ROP study [1] has evolved over a 15 year period (1974–1989). Each modification was based on our increasing knowledge of events triggering and perpetuating the disease. We conceptualize that ROP involves angiogenic stimulation emanating from spindle cells [2] and tractional forces resulting from myofibroblasts [3]. Both of these cell types reside on the vitreous side of the retina.

There are three objectives of transretinal cryotherapy which should be done sequentially at intervals of four to seven days. The first is to destroy the peripheral avascular retina from just anterior to the shunt to the ora serrata in order to obliterate all existing spindle cells. The second is to destroy extraretinal fibrovascular proliferation (EFP) and the shunt in order to obliterate myofibroblasts and their site of origin. The third is to destroy
Fig. 1. These two complete retinal montages demonstrate the macular distortion and ectopia which can result from cryotherapy with a therapeutic scleral buckle for Zone I ROP to the right eye of a 490 gram birth weight infant. Left: Preoperative appearance at the age of eight weeks with 1.3 cm of vascularized retina in the horizontal meridian through the optic disc and macula (distance between the pair of three vertical dashes), and with 0.4 cm between the temporal rim of the optic disc (two vertical dashes) and the center of the macular (circle). Cryotherapy was done at this time, two weeks later, and two weeks later. The retina totally detached between the second and third surgeries, and a therapeutic scleral buckle was placed during the third surgery. Right: Postoperative appearance at the age of 23 months with a therapeutic scleral buckle in place (anterior extent delineated by the wide solid circular line), and with 0.5 cm between the temporal rim of the optic disc and the center of the macula.

the connections between the central vascularized retina and the tractional remnants of EFP in order to create a new complete ora serrata.

A prophylactic scleral buckle should be applied concomitantly with the cryotherapy in order to keep the developing retina in constant contact with the choroid and sclera and to prevent the developing macula from distortion and ectopia. The rationale is to avoid rapid death of the immature retina by transient retinal detachment and to prevent macular damage related to photoreceptor misalignment by traction during three critical events: while transretinal cryotherapy creates multiple atrophic holes which coalesce to form a complete new ora serrata (one to two months following cryotherapy) [4], while remnant myofibrofibroblasts contract (two to four months following cryotherapy) [5], and while anterior rapid growth continues (four to six months following cryotherapy) [6].

Within our database, success is defined as a macula which always remains anatomically attached, but which may be distorted and ectopic. Fifteen years ago, our protocol was cryotherapy to the avascular retina alone. A recent review of our Zone I cases from 1974 to 1985 revealed only three eyes in two patients which were treated successfully by this technique (3/23 eyes in 13 patients, 13% success; 684 gram mean birth weight) [7, Table 17-1,