SYNTONOGRAM BULLETIN

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FLASHES FLASHES FLASHES

HOW OFTEN DO WE ASK, "WHY"

"Syndrome of Blue Sclera"

Well, we may not be interested in the question but we are vitally interested in "THE ANSWER," so let's try to see if the following quotations will assist us to understand more fully the reason "WHY" we use Alpha Delta or Alpha Theta in cases with "Blue Sclera."

From page 22, 1939 Year Book of Eye, Ear, Nose and Throat, the Year Book Publishers, Chicago, Illinois, I quote: "Syndrome of Blue Sclera - -Van Dar Hoeve's Syndrome. J. S. Charamis studied members of four generations of a family in which the syndrome of blue sclera, fragile bones and deafness was totally or partially present in a large proportion of each generation. The syndrome is familial and hereditary, of the primary mendelian type.

This family for the most part presented a type characterized by small bodies associated with large heads, often scoliosis or kyphosis, a ninepin-shaped thorax, and bowlegs with saber-sharp shanks. The greater number had repeated fracturs. Poor, irregular teeth with early caries, and early loss of teeth were common. Baldness was often present. The trunk was usually small in comparison with the extremities. Exophthalmos was present in a great number, and arcus Senilis developed relatively early in many. Blue sclera was a common finding. In certain number of loss of conjunctival structure was present. The author noted that the blue color of the sclera varied considerably in intensity in different persons. Anemia of the conjunctive, alternations in the choroid, diffuse cataract, strabismus, glaucoma and exophthalmos were also observed. The author believes that the most plausible etiology is disturbed metabolism of calcium and phosphorus due to defective functioning of the thyroid, parathyroids and adrenal glands. Treatment with hormones was unsatisfactory, but ultraviolet irradiation and raying the parathyroid glands combined with calcium therapy gave good results."

In the 1940 Year Book, which came off the press recently, the subject on Blue Sclera again attracts my attention. Page 24 "Blue Sclera. H.L. Jensen and K.K. Ortmann report two cases of Lobstein-Eddow's disease osteopathyrosis idiopathic. They state that with blue sclera there is a tendency to fractures and otosclerosis, constitution a triad. As in their second case, blue scleras are not always accompanied by the tendency to fractures. There is usually a relationship between the degree of coloring this tendency to fracture. All authors agree that blue scleras are the dominating symptom. The color has been observed in five generations and established in 50 descendants of one person. A certain intensity of the coloring, a familial characteristic and the combination of osseous and auditory stigmas in at least one member of the family are necessary for a positive diagnosis. The abnormal fragility of the bones depends on insufficient periosteal ossification with inability of the bone tissue to absorb calcium. The patients may become greatly deformed; in extreme cases they are known as "men of glass". The cause of the symptom complex is unknown. In one family the disorder appeared only in 20 males. Usually both men and women are affected. Transmission, to reappear as the result of disease - -exogenic or endogenic- -factors."

The above contributions to Ocular Literature is of vital importance. It is also very interesting to the Optometric Practitioner who has studied the Syntonic Principle, taken the Basic and advanced work.

How many of us remember that Spitler taught the importance of accurate "PRESCRIBINGSHIP" for a patient with "Blue Sclera" as long ago as 1929?

TO BE CONTINUED NEXT MONTH.

T. A. W. Elmgren, President
Columbia, S. C.

APRIL 1941

Now on with the subject continued from last month's Syntonogram. From the 1940 Eye, Ear, Nose and Throat Year Book I quote as follows:

"Syndrome of Blue Scleras, Fragile Bones and Poor Hearing- -Adair-Dighton's Syndrome- -. The syndromes of blue scleras, fragile bones, and poor hearing is also called Van der Hoeve's syndrome. Heinrich Rose describes three families presenting the syndrome, and presents Adair-Dighton's original case.

"Blue scleras were the commonest finding in the cases of partial syndrome. In a typical statistical study, - - Kuniis- -, blue scleras were found in 50 per cent of the cases, thinning of the bones in 45 percent and disturbances of hearing in 18 per cent. Other statistics vary. Their hereditary tendency is amply demonstrated.

"The blue of the sclera is variously described as azure-, porcelain and sky blue. It may involve the entire sclera or be entirely pericorneal or irregularly spotted. The difficulty with hearing is due either to otosclerosis or to labyrinth malfunction.

"Many other symptoms and findings have been described in patients presenting the whole or partial syndrome. Sixty-five were found by extensive perusal of the literature. It is difficult to classify this variety of defects.

"The etiology is not known. Pathologic studies reveal hypoplasia of the spongiosa and cortex of bone and calcium deposition around vessels. The scleras are frequently, but not universally, found to be one-half to one-third normal thickness. The bones are generally less radiopaque than normal owing to general osteoporosis. Normal blood calcium values as well as hyper-and hypo-calcemia are reported. The same variations are true of blood phosphorus values. Thyroid, hypo-physical, ovarian and pancreatic disturbances are reported. All races are affected. The great variety of findings has led to an equally great variety of therapeutic attempts.

"The syndrome is transmitted as a dominant hereditary trait."

This concludes the latest information available on "Syndrome of Blue Sclera". The information revealed is a of great value, which should warn the Ocular Practitioner against attempting to relieve ocular

dysfunction in Blue Sclera patients without a complete family case history. Many of the so called “grief cases” come from the Blue Sclera groups, therefore, we should be “doubly sure” that we are right before we attempt to relieve ocular dysfunctions in this group of patients.

So----------long,

T. A. W. Elmgren, Present