

EXOPHTHALMOS*

A. D. RUEDEMANN, M.D.

Exophthalmos or proptosis is a common condition of the eye which is present in numerous general as well as in many local conditions, and its presence constitutes a valuable diagnostic sign. It requires careful study and serious consideration because it produces disturbing functional and organic changes and also a distressing cosmetic effect. Although much has been written on various special divisions of this subject, little attempt has been made to correlate the various types and to enumerate their essential differences.

The study of exophthalmos in a large number of patients with thyroid disease has given me a keen interest in the problems presented by this condition, and, in order to evaluate the findings, the entire subject has been reviewed. The following notes are the result of a study of the patients we have seen, plus a review of the literature.

The total volume of the orbit is approximately 30 cc. and the eyeball occupies one-fifth of the orbital space. The eye does not lie in the center of the orbit, but a little to the side of the midline and is near the base of the bony cone anteriorly-posteriorly. It does lie in the center of the vertical plane.

The eyeball moves in all directions and can be moved anteriorly, posteriorly, and, to a slight degree, vertically. Several instances of persons who have well established voluntary propulsion have been reported. The eye is raised, with the help of the superior rectus muscle, approximately 1 mm. on extreme elevation of the lid. It can be pushed forward in the socket by leaning forward, holding the breath, or by compression of the carotid artery. The apex of the cornea does not protrude beyond the superior rim of the orbit in normal persons. The cornea and the anterior orbital fascia form the base of the outer or bony cone; the eye forms the base for the inner or muscle cone. The orbital contents, with the eye, fill the entire space in the socket, and any change in cubical contents is bound to alter the position of the globe. A diminution in space by compression from without or an increase in contents by an increase in the orbital tissue results in protrusion, whereas a loss of tissue due to atrophy or an increase in space by removal of a wall results in the exophthalmos which is so commonly associated with trauma.

The orbit contains muscles, vessels, glands, and nerves, and also a considerable quantity of fat. These elements remain fairly constant in

*This is the first of a series of four papers on exophthalmos and protrusion of the eyeball. The remaining papers will be published in subsequent issues of the Quarterly.

EXOPHTHALMOS

any one person, except in advanced age when a loss of fat allows a definite sinking of the globe.

The lack of valves in the outflowing veins allows venous congestion to take place easily and rapidly in case of any increase in intra-orbital pressure. The lack of space for expansion, by virtue of the bony lateral walls, the association of the surrounding sinuses with thin walls, the brain cavity above and posteriorly, and the numerous other structural relationships make malposition of the eyeball a fairly common and important sign of disease in adjacent areas.

METHODS OF MEASUREMENT OF EXOPHTHALMOS

The position of the eye should be estimated instrumentally and should be recorded for each individual case. Much effort and thought has been expended in trying to develop a universal exophthalmometer, but the variable factors exceed the constant factors so that relative rather than absolute values must be recorded and hence no definitely arbitrary limits of normal can be established.

Numerous instruments have been devised for the measurement of exophthalmos. A rough estimate of the degree of exophthalmos can be obtained by placing a ruler vertically from the brow to the cheek. This will not touch the cornea of an eye in the normal position, except in cases of myopia, and fairly accurate estimates are secured by this method. A testing frame with a flat lens can also be used, as can the ophthalmometer, by fixing the position of the head in the rest and measuring the necessary anterior-posterior movement of the mire case for each eye.

Instruments have been devised by Herring-Sattler which have been modified by Birch-Hirschfeld, and a more recent one by Kryosawa is patterned after a pelvimeter. The measurement is taken from the occipital protuberance and the tip of the other arm of the instrument is placed at the corneal apex. The objections to this method are the same as those to other instruments; namely, the individual variation in the structure of skulls, anteriorly-posteriorly, is too great to make possible an exact definition of the normal measurements.

In my studies, Hertel's exophthalmometer was used because it is simple to operate, relatively inexpensive, requires little time, is as accurate as any other instrument, and can be used repeatedly without replacement of parts.

It is my contention that each ophthalmologist should establish relatively normal values for the patients he examines, because the predominant race in any territory will aid in establishing the upper and lower limits of normal. After these have been established, he should then record the findings in each case. To measure the patient's eyes, the

vision should be fixed on the forehead of the examiner. Then, using equal pressure on both sides, one can accurately record the readings for any individual patient. The measurements thus obtained are of diagnostic significance. (In my own series of cases, I have measured each patient's eyes myself, so that the variable factor of individual interpretation has been eliminated.)

I have used ten groups of 100 patients each of so-called normals, consisting of cases for refraction, etc., and have established the following normal values for my own series: average values, O.D. 18.9 mm., and O.S. 18.7 mm., with a high normal of 20 mm. and a low of 14 mm. In some instances, the measurements have fallen below the scale of the instrument, and in some cases of exophthalmos, they have been beyond the upper limit of the scale, which is 33 mm.

Measurement of the width of the palpebral fissures is also useful. Here the variables are not so numerous or so great as in the anterior-posterior position. In the same series of normals, a ruler held across the middle of the cornea, with the patient's vision fixed on a distant object, yielded a measurement of 9 mm. O.U. Any exposure of the sclera above and below the cornea usually arouses suspicion that there is some protrusion of the eye or widening of the fissures. This measurement is important because many cases of so-called exophthalmos, especially "photographic" exophthalmos, are found to be merely a widening of the palpebral fissure. Hence a photograph without specific measurements is valueless in the diagnosis of exophthalmos. Relative ptosis, such as is seen in some instances of thyroid disease and in some patients with exophthalmos, makes it advisable to record the fissure measurement.

A measurement of the size of the pupil should be included in the study of each patient, as this aids in the diagnosis of certain other conditions and can be made easily when the other measurements are being recorded.

Visual estimation of the amount of proptosis is more inaccurate than is the finger tension in cases of glaucoma. The mere statement that the patient has exophthalmos without any record of the measurements is likely to be misleading and has caused, and is still causing, confusion, especially in the selection of cases for thyroid operations.

DIAGNOSIS OF EXOPHTHALMOS

A patient presenting signs of exophthalmos requires a thorough examination. I have established the following routine procedure in making the diagnosis in these cases.

1. Careful history, especially as to familial characteristics of eye position (particularly in patients with hyperthyroidism), and information concerning onset and duration of symptoms.

EXOPHTHALMOS

2. Vision—external examination.
3. Measurements—exophthalmometer, fissures, pupils.
4. Mobility of globe—diplopia, muscle measurements.
5. Refraction—ophthalmoscopic examination.
6. Intra-ocular tension.
7. Examination of fields (if indicated).
8. Location of pain and tenderness.
9. Temperature, blood counts including differential counts, urinalysis, Wassermann reaction.
10. Roentgenograms of orbit, sinuses, optic canal, and skull.
11. Physical examination in all cases of bilateral exophthalmos and in all but exceptional cases of unilateral exophthalmos including examination of nose and throat and determination of basal metabolic rate, etc.
12. Photographs of eyes, with measurements.

CLASSIFICATION OF EXOPHTHALMOS

The cases are divided into two main groups, (1) bilateral and (2) unilateral exophthalmos. After a careful study of my series of cases, and fairly diligent search of the literature, I have been able to list the following diseases of which exophthalmos is a sign.

BILATERAL EXOPHTHALMOS

- A. In children
 - I. Hemorrhage
 - a. Newborn, trauma
 - b. Spontaneous, as in hemophilia and blood dyscrasias
 - c. Scurvy
 - II. Increase in orbital content
 - a. Hemangioma, tumor, etc., (rarely bilateral)
 - b. Leukemia, chloroma, lymphatic edema, etc.
 - c. Xanthomatosis (Schüller-Christian disease and diabetes insipidus)
 - d. Tenonitis
 1. Serous
 2. Purulent
 - e. Endocrine disorders
 1. Hyperthyroidism
 2. Hypothyroidism
 - III. Decrease in size of orbit
 - a. Tower skull
 - b. Hydrocephalus

B. In adults

I. Hemorrhages

- a. Spontaneous leukemic, arteriosclerotic
- b. Traumatic
- c. Asphyxia
- d. Scurvy

II. Increase in orbital content

1. New tissue
 - a. Leukemia
 1. Mikulicz's disease
 2. Chronic lymphoma
 3. Lymphoblastoma
 - b. Bilateral tumor masses
2. Edema, venous stasis, etc.
 - a. Trichinosis
 - b. Tenonitis
 1. Serous
 2. Purulent
 - c. Cavernous sinus thrombosis and stasis
 - d. Hyperthyroidism
 - e. Malignant exophthalmos
 - f. Hypothyroidism
 - g. Hypertension
 - h. Allergic conditions
 - i. Arteriovenous aneurysm (orbital and cerebral)

III. Decrease in size of bony vault of the orbit

- a. Paget's disease
- b. Leontiasis ossea
- c. Acromegaly

IV. Relaxation of orbital fascia and muscle paralysis

- a. External ophthalmoplegia

UNILATERAL EXOPHTHALMOS

A. In children

I. Hemorrhage

- a. Newborn, trauma
- b. Spontaneous
- c. Scurvy

II. Increase in orbital contents

1. Inflammatory
 - a. Panophthalmitis
 - b. Orbital cellulitis (sinus disease)
 - c. Pyocele, pyemic abscess

EXOPHTHALMOS

- d. Cavernous sinus thrombosis; lateral sinus thrombosis
 - e. Periostosis, osteitis
 - f. Tenonitis
 - 1. Traumatic—surgical, foreign bodies
 - 2. Infectious
 - g. Foreign bodies
 - 2. Non-inflammatory
 - a. Intermittent (vascular)
 - b. Encephalocele
 - c. Meningocele
 - d. Dermoid cyst—teratoma
 - e. New growths
 - 1. Sarcoma
 - 2. Hemangioma
 - 3. Mucocele
 - f. Brain tumor
- B. In adults
- I. Hemorrhage
 - a. Spontaneous—leukemic, arteriosclerotic
 - b. Asphyxia
 - c. Scurvy
 - II. Increase in orbital content
 - 1. Inflammatory
 - a. Cavernous sinus thrombosis; lateral sinus thrombosis
 - b. Orbital cellulitis
 - c. Pyocele
 - d. Periostitis
 - e. Tenonitis
 - 1. Serous
 - 2. Purulent
 - f. Panophthalmitis
 - g. Foreign bodies
 - h. Granuloma—actinomycosis, mycelium, blastomycosis
 - i. Hydatid or echinococcus cyst
 - j. Pseudotumors
 - k. Tumors
 - 1. Gumma
 - 2. Tuberculoma
 - 3. Carcinoma
 - 2. Non-inflammatory
 - a. Air
 - b. Allergic conditions
 - c. Osteoma

A. D. RUEDEMANN

- d. Pulsating exophthalmos (may also be inflammatory)
 - 1. Trauma (77 per cent)
 - 2. Spontaneous (23 per cent)
 - e. Intracranial aneurysm
- III. Decrease in size of orbit
- a. Trauma
 - b. Hyperostosis, exostosis
 - c. Paget's disease

BILATERAL EXOPHTHALMOS IN CHILDREN

In the study of bilateral exophthalmos in children, several interesting facts present themselves. The first three diseases outlined—the hemorrhagic blood dyscrasias and scurvy—produce intra-orbital hemorrhages. The next four—hemangioma, leukemia, xanthomatosis and tenonitis—produce new tissue. In three of these—chloroma, xanthomatosis and lymphatic edema—the new tissue is formed directly from the blood elements. The tumors which cause exophthalmos are often hemangiomas, but any type, such as sarcoma, fibroma, and those of neurogenous origin may be found. When exophthalmos in children is due to an endocrine disturbance, the onset usually is later than in the other diseases listed, although the age of onset for various conditions cannot be set definitely. In either hyperthyroidism or hypothyroidism, the major problem is in the thyroid gland, but the general glandular system is severely involved; also the last two groups, tower skull and hydrocephalus, are definitely associated with physical changes in the skull which produce exophthalmos by making the orbit saucer-shaped, and by decreasing its volume content.

HEMORRHAGE

Trauma in the newborn: Bilateral exophthalmos in the newborn, due to hemorrhage, is rare, but several cases have been reported. Most instances of proptosis caused by hemorrhage are unilateral, but the possibility of this condition must be kept in mind also when both eyes are affected. Instrumentation, the use of forceps, or a prolonged delivery, especially in a primipara, may produce sufficient orbital hemorrhages in the infant to push the eye forward. These hemorrhages tend to clear rather rapidly, although the frequently associated cerebral disturbances which are evidenced by convulsions, etc., may possibly produce blindness, weakness, or paralysis. The ocular signs are transitory, the history usually is definite, and the diagnosis is not difficult.

Hemophilia and other blood dyscrasias: I have never observed a case of bilateral exophthalmos in a patient with hemophilia, but several such instances have been reported in the literature, and hence attention must

EXOPHTHALMOS

be called to this possibility in differential diagnosis. This blood dyscrasia, which is transmitted by the mother, is confined to male children only, and they may suffer severe hemorrhage from any trauma, however slight. The oozing usually is prolonged and of capillary origin. Cuts and lacerations generally are the precipitating cause of hemorrhage in these patients, contusions are next in importance, and spontaneous hemorrhages are rare. The history usually suffices to establish the diagnosis. Either eye may be affected by exophthalmos due to hemorrhage, but it is rare that both are involved at the same time.

Hemorrhages due to leukemia rarely produce bilateral exophthalmos, although the possibility must be taken into consideration. The diagnosis can be made from the blood picture. Examination of the fundus reveals engorgement of the veins and, at times, hemorrhages.

Scurvy: One of the causes of exophthalmos early in life is scurvy which is due to lack of vitamin C. This deficiency disease appears when there is a lack of breast milk, usually as the baby is being weaned, and before the proper formula for feeding has been determined. Naturally it is most prevalent in families afflicted with poverty or ignorance. Children less than one year old are most frequently affected, although instances in older children have been reported, especially in times of depression and among the poorer classes.

Associated with the proptosis and of even more frequent occurrence in scurvy are disturbances in the epiphyseal junctions of the bones of the legs. Early flexion of the legs is followed by eversion and immobility, and finally, by a state of pseudoparalysis. Diarrhea is also an accompanying symptom.

The pathologic basis for the hemorrhage in these patients presumedly is a lack of cement substance in the endothelial cells of the blood vessels, which makes them extremely permeable. The seat of the hemorrhage producing the exophthalmos usually is between the orbital plate and the periosteum. The hemorrhage invades the upper lid, and comes on suddenly following a trifling injury or a fit of coughing or crying. There is a statement in the American Encyclopedia of Ophthalmology that a black or protruding eye which occurs during the first dentition and is not due to injury is more likely to be due to scurvy than anything else.

The eyeball, usually on the left side, is pushed downward, forward, and laterally. The amount of proptosis is slight, whereas the lid ecchymosis may be great. The other eye may not be involved for several days or until after the condition of the first eye has begun to improve. No local orbital treatment is advisable because pressure and manipulation produce additional hemorrhage. Rest and the proper diet are the essential factors in treatment. The antiscorbutic diet, which includes

large quantities of lemon juice or orange juice, begins to effect recovery within four to five days. However, the treatment must be prolonged, or additional hemorrhages may occur.

INCREASE IN ORBITAL CONTENT

Tumors, hemangiomas, etc.: The tumorous conditions which produce bilateral increase in the orbital content are not numerous, and new growths are rarely found simultaneously on both sides. However, a few such instances have been reported, the diagnosis usually being made at the time of operation. Multiple dermoid cysts or hemangiomas might be suspected in cases of bizarre bilateral exophthalmos. In such conditions, the proptosis would not be typical of the bilateral exophthalmos seen in other diseases.

Leukemia, chloroma, lymphatic edema, etc.: Invasion of the orbit by overproduction of the blood elements fortunately is rare. The cases reported and the three or four that I have seen all have terminated fatally. Probably the most common and also the most spectacular growth of this type is the chloroma which is seen in myelogenous leukemia. It derives its name from the greenish coloration of the skin and tumor mass. Much conjecture regarding the etiology of these growths and their true classification is found in the literature. They probably have their origin in an acute leukemia with a predilection for the periosteum, especially in the cranium. The leukemia is of malignant type and the cells apparently have the power to multiply in the blood stream and tissue spaces. Various forms of cells are described. These range from the large lymphocytic and atypical forms to the aleukemic forms, which are the more malignant. The anemia is due to the crowding out and replacement of the erythrocytic blood elements and lessening of red cell formation. The malignancy of the tumor depends on this replacement factor.

The points of interest to the ophthalmologist are that the exophthalmos usually commences in one eye, with rapid subsequent involvement of the other orbit. The invasion by the tumor may be posteriorly or from above. The mass is made up of a greenish tissue substance, which fades rapidly on exposure to air, supposedly as the result of the oxidation of the bilirubin it contains. Accompanying signs and symptoms are weakness, anemia, headaches, tinnitus, deafness, and changes in the blood indicating leukemia.

The blood picture shows a red count in the neighborhood of 3,000,000 and hemoglobin from 70 to 80 per cent. The differential count shows an increase in the mononuclear leukocytes and in the large and small lymphocytes.

EXOPHTHALMOS

Removal of the tumor masses and treatment by radium and roentgen radiation are merely palliative. No cures have been reported in this type of case.

Other forms of leukemic tumors have been described as causing exophthalmos. Major Wright reported a case of lymphoblastoma in which there was an increase in the mononuclear cells (monocytes and transitional cells). The patient, a child twelve years of age, had bilateral orbital growths, although only the left eye was proptosed. The boy died within a short period after he was observed.

The so-called lymphomas may produce bilateral protrusion of the eyeball, and diagnosis is difficult because of lack of signs and symptoms of systemic disease. The blood picture may not be strikingly abnormal, and, unless the tumor is sectioned, evidence of the true diagnosis will be entirely lacking. In this type of case, it often is necessary to have blood counts made repeatedly, for the cells may vary from time to time and several examinations may be required to reveal an increase in white cells.

Xanthomatosis (Schüller-Christian disease), Lipogranuloma: The rarity and uncertainty of knowledge concerning a disease may sometimes be estimated by the number of titles attached to it. This disease, first designated as xanthoma by Rages in 1836, has been described repeatedly since then. Excellent discussions of the subject by Wheeler, Knapp, Heath, and Rogers are to be found in the literature on ophthalmology. Several subdivisions of this condition, such as Gaucher's disease and Niemann-Pick disease, are grouped under lipid dystrophies but these are not accompanied by exophthalmos.

Exophthalmos due to xanthomatosis is becoming more familiar to oculists and is being diagnosed more frequently; hence it may be expected that more descriptions of cases of this type will be recorded in the literature. Personally, I have seen two cases, one in a young girl, and one in a young boy. The disease occurs most frequently in children; only six of the fifty-five patients reported in the literature were more than twenty years of age. Remissions and relapses characterize the condition and new points of invasion may be evident at any time.

The cases reported represent all types of this disease, according to the parts involved. The eyeballs usually are pushed straight out by a mass of fibrofatty tissue in the orbit. This tissue is lipogranuloma, made up of three major portions, foamy lipid cells, inflammatory cellular exudate due to tissue reaction, and fibroblastic proliferative tissue.

There may be map-like changes in the skull, due to a pressure atrophy by the histiocytes. In some patients, the pituitary gland may be involved in the process with resultant diabetes insipidus. Extreme thirst and fre-

quent micturition, a sore mouth, loose teeth, dwarfism, amenorrhea, pathologic fracture, discharge from the ear, anemia, and shortness of breath constitute signs and symptoms of this condition. Many variations are noted in individual cases, and neither exophthalmos nor diabetes insipidus are essential to the diagnosis of xanthoma.

The disease is caused by faulty lipid metabolism and little is known of its etiology. About 30 per cent of all cases terminate fatally. Roentgen irradiation associated with a diet low in fats and plenty of exercise to increase metabolism has offered the best therapeutic results. Defects in the bone may completely heal following this regimen.

Bilateral tenonitis: Bilateral serous tenonitis is a rare condition producing but slight degrees of exophthalmos, although the disturbances in the orbit are severe. The discomfort of the patient is out of all proportion to the extent of the edema in the anterior portion of the orbit. The process usually begins in one eye and may be present merely over one small area. The conjunctiva is edematous and pale yellow in color. Movement of the globe causes pain. As the disease progresses the patient may have a slight fever. A little later, the other eye may become involved and the entire course of the disease may last from several days to a week. The eyes are exquisitely sensitive and the patient refuses to move the eyeballs.

Treatment consists of the use of salicylates, heat, and general supportive measures. The prognosis is good.

I have seen one patient with bilateral tenonitis which was due to diabetes and the eye condition cleared promptly when the patient became sugar-free.

Purulent tenonitis usually is unilateral following injury, although I have seen one patient in whom the process was bilateral following tenotomy of one eye. Extreme exophthalmos developed which resulted in some loss of sight in both eyes.

Endocrine disorders (Hyperthyroidism, hypothyroidism): Bilateral exophthalmos in children does not occur frequently, and, in my own experience, endocrine disturbances have accounted for most of the cases. These conditions are described fully under the classification of exophthalmos in endocrine diseases in adults, and hence no detailed discussion will be presented here. I shall merely mention that in nervous, irritable children who are not gaining weight, and who show signs of widening of the fissures with slight protrusion of the eye, hyperthyroidism must be considered. On the other hand, those who gain weight easily, are slow in motion and thought, and have heavy eyelids, may well belong in the group with hypothyroidism.

EXOPHTHALMOS

Both these groups of patients present interesting problems to the ophthalmologist. For the treatment of instability of the ocular muscles, exercises frequently are ordered which prove of no value. This muscle instability varies from day to day and must be central nuclear in origin rather than peripheral. Some of these children are referred by school authorities for eye examination, especially for the fitting of glasses. The frequency with which glasses are obtained is a good indication of the general lack of knowledge in regard to diseases of this type in children. In our series of cases of exophthalmos due to thyroid disease in both adults and children, 75 per cent were wearing glasses at the time of examination. These had been prescribed during the course of the disease, and some patients had even received several pairs, while the exophthalmos continued to progress.

Protrusion of the eyes may be just as marked and just as serious in children as in adults, and it is important to realize that although some of these children require operations on the thyroid gland, others may be benefited by rest in bed and general measures, including glandular therapy. Great care and judgment must be exercised in the treatment of these conditions, for many of the general symptoms may be transitory. The unwise use of glandular extracts and other therapeutic measures may precipitate more serious difficulties in these patients. Thyroid extract and the new pituitary products should be prescribed only by those with considerable experience in clinical endocrinology.

DECREASE IN THE SIZE OF THE ORBIT

Tower skull: A variety of congenital malformations of the skull may involve the orbit. In some instances, the deformity is slight and in others extreme, with the head pointed and the eyes almost luxated. This is accompanied by loss of vision, cerebral deficiency, and other associated signs and symptoms which include optic atrophy, nystagmus, headaches, convulsions, and hyperopia.

The disease is not commonly reported to cause exophthalmos but this is because of lack of recognition rather than lack of frequency. Since the excellent report by Davis in 1925, eight patients have presented themselves at the Cleveland Clinic with exophthalmos which might be classified as resulting from this malformation.

The two forms most often described are the dome-shaped, tall head, and the extremely high, pointed type. Schüller mentions three forms, the tower skull, the short, broad head, and the abnormally high, pointed type. Exophthalmos, although rather rare, has been reported to accompany all three types; frequently in the first, and less often in the second and third groups. One reason for this is that most authorities classify

all these deformities under the first group, tower skull, and do not refine their definitions.

The etiology of these malformations is unknown. Syphilis was not the cause in any of our eight cases nor in the six that Davis described. Some fault in development produces the deformities these patients display, as other congenital anomalies, such as webbed fingers and toes, cleft palate, medullated nerve fibers, etc., often are present. Males are more frequently affected than females. The mental disturbances are probably not due to stenosis of the brain, but rather to a combination of malformation of the brain and lack of development of the skull. Wiggers maintains that the bone grows around the tissue and fits it, and that the tissue does not expand to fit the bone.

A patient with hyperopia, secondary optic atrophy, and nystagmus may have a mild form of this disease, and the exophthalmos may vary from that of slight degree to actual luxation. This suggests the vast number of types and combinations of eye symptoms and signs which may be present in these patients.

The orbit becomes saucer-shaped, the roof is almost vertical and the lesser wing of the sphenoid is vertical rather than horizontal. The orbit may be further shortened by pushing forward of the greater wing of the sphenoid. The entire picture is one of a short orbit with insufficient room for its contents. Nystagmus is of the visual type and may have its origin in congenital amblyopia or may be due to visual deficiency of postneuritic or hyperopic origin.

Cranial deformities revealed by roentgenographic examination are numerous and include (1) the high-domed, pointed skull, (2) the severe form with a short anterior-posterior diameter, (3) wide, bulging, temporal fossae, (4) shallow orbits, (5) high, narrow palate, (6) deviation of the nasal septum, (7) facial asymmetry, and (8) honeycombed skull due to convolitional markings and cerebral pressure.

The malformation may cause headaches due to the cerebral stenosis or to the visual defect. These patients also have muscle and refractive errors. Convulsions were not seen in any of our patients, and are rare.

Treatment should be directed toward the cerebral cavity. Decompression, if performed early, may benefit the patient, although death following operations is not unusual in these cases. Fenton has made a plea that these patients be treated early and he believes that radical measures are justified, because of the patient's extreme hideousness and the progressive cerebral decline and continued visual loss.

Hydrocephalus: Hydrocephalus is encountered quite frequently in a diagnostic clinic where there is a neurologic surgeon, and I have seen numerous cases of both internal and external hydrocephalus. Exoph-

EXOPHTHALMOS

thalmos is not a major sign in this condition, and is seen but rarely. When present, it usually is bilateral and is produced by a pushing forward of the floor of the orbit. In contrast to tower skull, the sutures in hydrocephalus may be open, and this, although allowing tremendous enlargement of the skull, protects the orbit.

Associated with the enlargement of the head may be edema of the discs with either primary or secondary atrophy of the optic nerve.

Roentgenograms reveal the relative increase in size of the various parts of the cranium and ventriculography reveals the extent of the fluid increase. Various treatments have been devised for this condition, and at present it appears that several of these methods offer considerable promise.

Summary: In considering the entire group of children with bilateral exophthalmos, roentgen examination is of value in four, namely scurvy, xanthomatosis, tower skull, and hydrocephalus. Blood studies yield important information in scurvy, the leukemias (chloromata) and possibly in xanthoma, if the cholesterol is increased. The basal metabolic rate is of importance in the diagnosis of exophthalmos due to increased or decreased activity of the thyroid gland.

Almost all cases of bilateral exophthalmos are systemic in origin and hence a careful physical examination may reveal the diagnostic clue.