EXOPHTHALMOS*

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UNILATERAL EXOPHTHALMOS IN CHILDREN

Pyocele: The infection in pyocele usually originates in one of the ethmoids although the antrum may be involved. These youngsters are quite ill, there is an increase in temperature, they have general malaise and complain of pain through the side of the head. Within a short time, signs of increasing congestion, edema of the lids, and some exophthalmos appear. The eye may not be pushed as far forward as it is laterally. There are no changes in the fundus early in the course of the disease but, as the inflammation proceeds, increasing congestion of the veins occurs and later the eye may show a low grade intra-ocular inflammation.

Examination of the nose usually reveals definite clinical evidence of sinusitis. Roentgen examination may show clouding of the sinuses but it rarely shows the orbital entrance or orbital lesion. Use of the suction apparatus is an aid in establishing the diagnosis.

In treating a patient with pyocele, he must first be put at complete rest with heat to the side of the face and eye. The nose must be kept open and constant drainage maintained by the use of suction or irrigation. The orbit should not be incised unless necessary as it is sometimes possible to avoid this procedure. Efforts should be directed toward causing the abscess to localize and, if necessary, the area or a dependent part of the orbit may be incised for drainage. Constant attention must be given to maintaining the integrity of the orbital contents in order to preserve the function. The nasal and supportive treatment must be continued for some time.

Pyemic abscess is a secondary process. The patients usually have had a general infectious disease which has lowered their resistance, and the orbit happens to become the seat of a metastatic infectious thrombosis. The associated exophthalmos is usually directed outward or inward, according to the location of the abscess. Since most of them are outside the muscle cone, the eyeball is not pushed straight out. Associated with this, there may be an increase in the white cell count and a slight elevation of the temperature.

Treatment consists of hot compresses, bed rest, and general measures. If the protrusion is progressive, then the orbit should be incised for drainage.

Orbital cellulitis is a serious inflammation of the retrobulbar tissue due to a foreign body, injury, a metastatic infection or direct extension

*Concluded.

from the sinuses. The lids become red, edematous, and swollen, and the conjunctiva is infected. There is exophthalmos and marked impairment of muscle function with deep boring pain in the orbit. This is a constant dull ache. The infection may be due to any of the usual group of organisms such as Streptococcus or Staphylococcus. Cases are reported in which the B. pyocyanus, typhoid bacillus or Pneumococcus was responsible. Early supportive treatment is essential and in spite of good therapy, 20 per cent of the patients with this condition lose the sight in the affected eye. An abscess may form, causing thrombosis of the central retinal vein, occlusion of the artery, or optic retinitis. The cornea may break down and the entire globe may degenerate in the course of the infection. Meningitis and cavernous sinus thrombosis result in a 17 per cent mortality.

Cavernous sinus thrombosis occurs very infrequently in children and will be discussed under exophthalmos in adults.

Lateral sinus thrombosis has been reported to cause unilateral exophthalmos secondarily. I have never seen exophthalmos but I have seen papilledema. The exophthalmos is a retrograde process and is caused by a general venous stasis which extends over the mastoid bone by way of the emissary veins. These patients are only benefited by treatment of the lateral sinus affection. In cases of papilledema and papillitis, the end result depends upon the time element, the shorter the duration of the involvement of the optic nerve, the better the visual result.

Periostitis: This is not a common disease of children and usually is found only in those suffering from malnutrition and general physical debility. The majority of cases are due to tuberculosis of the lateral wall of the orbit, the inferior outer rim being most frequently involved although the superior margin may be included. The characteristic picture is first hyperemia of the area and this is associated with some swelling and pain. The eyeball is rarely pushed out, usually being displaced inwardly or downward and inward. As the swelling increases, incision will effect drainage of some free pus and probing will lead to uncovered bone. General measures, such as heliotherapy, high calorie, high vitamin diet, and rest are the best measures.

Luetic periostitis (a tertiary leutic process) will produce inflammation, swelling, and pain which usually comes on at night. Luetic periostitis suppurates but rarely in contrast to other types of periostitis, and blood tests on the parents and child are of value in establishing the diagnosis.

Osteitis may be associated with the periostitis and is but rarely seen in the region of the orbit unless some complicating disturbance is present. Necrosis of the bone necessitates its removal and then treat-

ment must be instituted to build up the patient. Attention is called to the possibility in older children of osteomyelitis infiltrating from the frontal sinus into the orbital bones and throughout the walls of the orbit. I have seen several of these cases in which the removal of the bone did not cause much intra-orbital involvement except for congestion and a low grade exophthalmos with some disturbance of muscle balance. To effect a cure, one must be willing to be very radical in the excision of diseased bone; otherwise the process continues to extend.

Tenonitis: Inflammation of Tenon's capsule results in a low grade exophthalmos of only 1 or 2 mm. but the eye is very painful, especially on movement. Any injury to the orbit may institute a low grade tenonitis and penetrating wounds are apt to promote a localized inflammation in the capsule. Here the pain and swelling will be limited and movement of the eye will be painful, especially when the muscle in the area involved is stretched.

Surgical tenonitis subsequent to surgery of the ocular muscles may be very serious, very painful, and is certainly most annoying to the surgeon as well as the patient. This may be caused by irritation from the suture material or an infectious process. Occasionally, slough of the muscle tendon or an unusual amount of scarring will interfere with the desired surgical result.

Foreign bodies may enter the orbit and disappear in the orbital tissue. Small pieces of material usually are lost and, unless they produce signs or symptoms, it is better to do nothing about them. Larger bodies, such as sticks of wood, pieces of copper, and substances that produce an inflammatory reaction must be removed not because of the exophthalmos but because the secondary reaction may produce fixation of the muscles, diplopia, or deep cellulitis.

Noninflammatory

Spontaneous hemorrhage into the orbit is very rare but may occur in association with scurvy, leukemia, or any severe blood dyscrasia. Trauma at birth or later does not occur commonly and the protrusion in any event is not great. I recently saw a case in which severe exophthalmos occurred in malignant hypertension due to hemorrhage following rupture of the orbital portion of the ophthalmic artery.

A diagnosis of *intermittent (vascular) exophthalmos* due to varices of the orbital vessels, telangiectasis or vascular tumors such as angiomata or lymphangiomata is very difficult to establish. It is sometimes of value to have the patient bend over sharply so that there is increase in the venous stasis and a secondary increase in the exophthalmos. An increase in size may occur while straining, crying, and,

in some instances, during the menstrual period. These changes are slowly progressive and, when the eye is definitely moving forward, the tumor should be removed.

Encephalocele is a congenital defect which does not produce much exophthalmos as a rule, but it displaces the eyeball downward and outward. The bony dehiscence is between the ethmoid and frontal bones, and it is sometimes possible to demonstrate this by roentgen examination. The cyst is soft and may be pressed out. The mass may be replaced and the opening covered by a bony plate.

Meningocele is a congenital condition which is very rare. It occurs at the same location as an encephalocele, is more soft, and may pulsate. It must be differentiated from encephalocele and dermoid cyst. Roentgen examination and aspiration with a large bore needle will aid in the diagnosis. The cyst produces little or no exophthalmos but moves the eye downward and outward. Extreme care must be exercised in the management of these conditions because of the danger of meningitis.

Recently I saw a patient who had been in an accident and had a large cyst in the upper inner angle. There was some protrusion and considerable malposition of the globe. The preliminary diagnosis was pyocele extending inward from the anterior ethmoids. Dr. Paul M. Moore of the Department of Otolaryngology proposed to exenerate these cells but fluid obtained through an incision showed the cyst to extend into the orbit. Further exploration and excision showed a cystic degeneration of the dome of the lachrymal sac which had been cut off at the time of the accident and displaced upward and inward. Removal resulted in complete recovery.

Dermoid cysts rarely produce exophthalmos as they are usually superficial, extending upward and outward, and although they may push the eye downward and inward, exophthalmos is not a common finding. They are congenital and contain exfoliated epithelium, hair, oil and rarely teeth. Removal is indicated for cosmetic reasons and because of the exophthalmos.

Teratoid cysts or teratomas occur but rarely and usually they contain more tissue than do the dermoids. They also are congenital and may continue to develop. Parts of a fetus have been found and a case is on record of one teratoid growth which was made up of an almost complete fetus. There are cysts due to old hemorrhages, and a rare cyst of the trochlear of the superior oblique.

Thyroid disease occurs in children and as noted in the discussion of bilateral exophthalmos, it produces exophthalmos fairly frequently. This usually is bilateral and any unilateral exophthalmos may be relative because of the unequal widening of the palpebral fissures.

The latter sign occurs earlier than true exophthalmos and disappears rapidly following treatment of the thyroid disease. Progressive exophthalmos as proved by measurements requires surgery of the thyroid gland. Care of the eyes should be taken to prevent ulceration due to exposure. The longer the exophthalmos has been present, the less recession may be obtained and the more difficult becomes the problem of reëstablishing normal ocular muscle balance and position of the eyes.

Tumors of the orbit in children are rare and of mixed types. Vascular tumors have been discussed under intermittent exophthalmos. Sarcomata, either round or spindle cell, occur and these either extend back along the nerve or in or around the nerve itself. Any connective tissue in the orbit may give rise to a sarcoma. These tumors may cause little or no exophthalmos for a long while, but they usually produce loss of vision early if they arise in the muscle cone. The exophthalmos, because the tumor is inside the muscle cone, is straight forward. The usual sarcoma is round, soft, and encapsulated, and may cause any type of protrusion.

Mucoceles are rare and because of their anlage in bone they invade the orbit. The exophthalmos usually is not straight forward.

Rarely are *brain tumors* allowed to progress so long that they extend into the orbit. Tumors of the anterior lobe or tumors in the middle fossa may obstruct the venous outflow and produce a secondary orbital edema.

Congenital malformations are apt to produce an exophthalmos. Maldevelopment of one side of the head may produce a relative exophthalmos on the unaffected side. An enophthalmic eye may cause the normal side to have the appearance of exophthalmos. The cases of "Türmshadel," "Spitzkopf" (oxycephaly) or "tower skull" may be associated with a serious type of unilateral exophthalmos. A prominent eye or eyes may be accompanied by many varying degrees of changes in the skull, and the appearance of the patient is apt to be grotesque. He is uncomfortable because of the muscle error and exposure and sees poorly because of the optic atrophy, so that he usually is an unhappy individual. Hydrocephalus, as a rule, does not produce exophthalmos.

Unilateral Exophthalmos in Adults

An adult is more prone to unilateral exophthalmos than is a youngster. The danger of trauma is greater, and because of his additional years, he has accumulated pathological processes and physical debilities which now begin to exact their toll.

Various blood dyscrasias may result in a *spontaneous hemorrhage* of the orbit which will produce an acute exophthalmos. This usually

is not very great, being 2 to 3 mm. and if the general process is controlled, a normal position of the eye and normal function may be recovered. However, the hemorrhage may not resolve very slowly and blindness may ensue or a blood cyst be formed.

Arteriovascular disease with or without hypertension may produce hemorrhages of the orbit which vary in size and extent. The resulting exophthalmos may be small or great according to the location of the hemorrhage and the amount of blood lost. Outside the muscle cone, the blood suffuses anteriorly and may appear in the lids and ocular conjunctiva. Hot compresses, bed rest, and a pressure bandage should be prescribed and general measures should be directed toward reducing the blood pressure.

Asphyxia, especially where there has been violent coughing and choking, may result in an orbital hemorrhage. This is usually associated with intra-ocular and conjunctival hemorrhages. Severe compressing injuries of the chest may also produce ocular hemorrhage. When crushing blows are received in which a heavy weight or pressure suddenly compresses the wall of the chest even though it may rebound without fracture, the exerted pressure may be transmitted throughout the vascular system and the vessels cannot compensate for the sudden mechanical pressure. The hemorrhages will be present in the retinae.

Scurvy and other deficiency diseases that change the blood volume may result in orbital hemorrhage. Today, scurvy in this country is very uncommon, although the possibility of its occurrence must be borne in mind, especially when dealing with adults who have decided how they should eat and with faddists who believe they know how to eat.

The *inflammatory processes* which cause increase in the orbital content are numerous. The orbit is well located for protection but unfortunately man insists on exposing himself to many undue risks. The cushioning effect of the air cells around the orbit are a boomerang when they become infected. In spite of this the orbit is only infrequently involved, considering the number of injuries to the head and infections of the sinuses. Probably more cases of slight exophthalmos are seen than are recorded, but evidently the eyes recover their normal function and position. It is only when noticeable and when exophthalmos is associated with pain and disturbances of function that the oculist is consulted as to the cause and effect of the existing condition. The infectious processes are the same as in children with some increase in the number of the following conditions.

Fortunately, the number of cases of *cavernous sinus thrombosis* is not great. It should be emphasized that an infection of the sinus may spread from a remote or localized infection and then produce the orbital congestion secondarily. The end result is usually the same but it is

difficult to determine the etiology at the onset. Lateral sinus thrombosis may also produce exophthalmos but this is by secondary edema. It is well to ascertain that this is not the true condition before the orbit is incised or explored. However, it is also well to know that the orbital lesion may be a pyocele or cellulitis before any attempt is made to drain a lateral or cavernous sinus.

Periostitis and osteitis are rather uncommon conditions of the orbit but I have seen cases in which there was a low grade, slowly progressive, painful type of exophthalmos. Although these diseases are usually thought to be due to lues the infections were nonspecific in my cases. Treatment is of little value but heat locally and deep therapy should be tried to afford relief of the pain.

Tenonitis as discussed under exophthalmos in children is similar in the adult except for one group of patients. In the serous or rheumatoid tenonitis associated with gout and general metabolic disorders, a greater number of cases of exophthalmos are recorded.

Fortunately, the protrusion is only 2 or 3 mm. The pain is out of proportion to the proptosis and is due to some involvement of the muscles. Rest, heat, and elimination of foci of infection are important. Heat by the cabinet or diathermy method may well be tried. Large doses of salicylates are beneficial and typhoid therapy is of value. The patient should have a specified dietary regimen.

The exophthalmos of *panophthalmitis* occurs late in the course of the disease and is slight but the pain again is out of proportion to and not due to the protrusion. The inflammatory process with the heavy cellular infiltration fills up the orbital space and the eye moves forward. In some instances, the entire globe is increased in size. Care is essential in handling these severe cases.

The method of handling *foreign bodies* in adults is the same as in children—those that are nonirritating are best left without treatment. The functional disturbance will probably not be as great as when one attempts too much surgical intervention.

An occasional case of infectious *granuloma* of the orbit has been reported, secondary inflammation being the cause of the exophthalmos. The infecting fungus may be actinomycosis, mycelium, or the blastomycosis. Draining sinuses from the orbit are produced and biopsy and careful bacteriological study are necessary to establish a diagnosis.

Hydatid or echinococcus cysts occur very uncommonly in the United States. The cyst is usually metastatic, the primary cyst being found in the liver.

Most *new growths* which produce exophthalmos are unilateral and noninflammatory. An increasing number of cases of inflammation of

the orbit are reported in which surgical intervention failed to reveal a tumor. Benedict drew attention to this some years ago and the name pseudotumor of the orbit has been given to this condition. In the presence of this inflammatory new growth, the eye and orbit are painful, the eye moves forward slowly, movement is limited and may be painful, and a gradual loss of vision takes place. Treatment by deep x-ray and local heat have been of little value. An associated infection of the sinus may be the etiological agent and careful study should be made in all cases. Exeneration may be required but the orbit continues to fill up with inflammatory tissue. True new growths of the orbit which produce inflammation and exophthalmos are rare although exophthalmos is the sign most common in all new growths. The displacement and the motility of the globe are important localizing signs. The progress of the protrusion, the associated pain, and its location should be taken into consideration.

Gummata are tertiary luetic manifestations which occur but rarely and they usually are unilateral. The positive blood test, the presence of scars from the primary lesion, or associated signs of lues should be looked for. The history and the onset of pain at night are aids in establishing the diagnosis. Trial therapeutic measures of neoarsphenamine or massive doses of potassium iodide may help to solve the problem and relieve the pain.

Tuberculoma are very rare and need only be mentioned as some cases are on record. They probably are the most difficult to diagnose preoperatively unless strong supporting evidence is present. As is true of tuberculosis in any part of the body, tuberculoma may break down and form a fistulous tract. Treatment should be directed toward general measures such as a high vitamin, high calorie diet, bed rest, and heliotherapy.

Exophthalmos from *carcinoma* is usually the result of metastasis or invasion. The glandular type arising from the lachrymal gland is fairly easy to diagnose and does not produce exophthalmos until late in the course of the disease. The eye is usually pushed downward and inward. Basal cell tumors of the lid should be treated early by radium or roentgenotherapy and then should be watched for recurrence or for new tumors in adjacent areas. Some of these tend to invade early and, although the invasion is inflammatory, it may be attributed to the previous radium or roentgenotherapy and the true nature overlooked. Here again exophthalmos is late and exeneration is necessary. Carcinoma which invade the orbit are quite common, usually coming from the antrum although the ethmoids and the sphenoids and rarely the frontal sinuses may be the seat of the original lesion.

These infiltrating carcinoma result in an early fixation of the globe with diplopia and a progressive type of exophthalmos. This is undoubtedly the most devastating of the new growths and requires radical methods of treatment. This condition is treated by radium or deep x-ray where possible. In other instances, radical exeneration and implantation of radium and use of x-ray are employed. It is advisable that patients who have had a malignancy of this type report at regular intervals over a period of time.

The *noninflammatory conditions* produce many cases of exophthalmos but are few in number.

The fracture of an orbital wall or loss of the wall due to syphilis or tuberculosis may result in the admittance of air into the tissues by way of the sinuses. Blowing the nose, sneezing, or coughing may push more air into the orbit and the exophthalmos may be sudden and serious. However, in most instances, pressure on the globe will usually meet with little resistance and a temporary pressure bandage may suffice to correct the condition. The patient must be warned about the cause of the condition and that care must be exercised or infection may be blown into the orbit by the same route.

Unilateral orbital edema due to *allergy* is rare but may occur. Unilateral edema due to congestion of the sinuses is seen. Shrinking of the nasal mucous membrane and irrigation of the sinus will aid in relieving the condition of the orbit and where drainage of a sinus is interfered with by a deviated septum or large boggy turbinate, the mechanical difficulty should be handled expeditiously. The instant there is involvement of the orbit, it is well to establish a diagnosis and institute treatment because any delay may mean a loss of vision.

Osteomata invading the orbit are not uncommon. They are produced by irritation of an associated sinus and may attain the size of a large robin's egg. Removal is necessary and the nose should be examined for any possible source of irritation. Many types of sarcoma invade the orbit and early removal is advisable. Most sarcomata in the orbit are not very inflammatory but form a fairly well isolated tumor mass which can be removed. The tumors arising from the orbital tissue do not, as a rule, metastasize early. Unless they are excised completely, they may recur. The method of approach is according to the location of the tumor. The lateral posterior route as advocated by Krönlein should not supersede other approaches unless one is relatively certain that the main tumor mass is well posterior and cannot be reached by easier and less traumatizing procedures.

Pulsating exophthalmos may be accompanied by visible pulsations which are also audible to the patient. It is fairly common and usually

unilateral. Certain features are common to both the traumatic and spontaneous types: (1) Both are acute; (2) the patient is conscious of the bruit; and (3) the exophthalmos is not great. Holloway reported a number of these cases and Locke, in 1924, collected a large series, giving an excellent description of the condition and the various forms of treatment. He found that the largest number of cases of pulsating exophthalmos were not spontaneous, but were due to trauma from injuries in automobile accidents. When this condition comes on, the patient is conscious that something has happened and there is an associated loss of vision which may produce complete blindness. Although there is some arterial circulation, the ischemia of the retinae is sufficient to cause loss of vision. The fall of arterial pressure results in a rise in the intra-ocular pressure and this also is probably a factor in disturbing the retinal circulation and producing a secondary anemia. Permanent changes in the retinal tissue may occur very soon because nervous elements do not tolerate lack of circulation over a very long period of time.

Locke described in detail the various types of treatment of pulsating exophthalmos by use of trial periods of compression. Gardner has employed muscle implants in the vessel, allowing them to drift to the site of rupture as advocated by Adson. This has met with considerable success. The condition, on the whole, is a serious one and the visual results, as a rule, are poor. Much of the exophthalmos remains because of the secondary fibrosis that takes place.

Intracranial aneurysms may cause exophthalmos because of remote congestion in the veins of the orbit. This is very unusual although Cushing and others have reported a number of cases. It is not easy to establish a diagnosis and the treatment must be directed by a neurosurgeon to the intracranial lesion.

A relative unilateral exophthalmos may occur in *thyroid disease*. We have not seen any cases in which the exophthalmos remained unilateral once the disease was established; however, cases have been reported by well qualified men and must be given consideration. It is safe to say, however, that given a case of unilateral exophthalmos with little substantiating evidence of thyroid disease, one should seek further for the cause and even in those cases where some thyroid disease is in evidence, other conditions should be carefully ruled out. The case in which widening of the palpebral fissures gives the appearance of exophthalmos is very misleading and, unless careful measurements are made, one is apt to make an incorrect diagnosis. Unequal exophthalmos is very misleading and, unless careful measurements are made, one is apt to make an incorrect diagnosis. It is also very common and because the measurements are of most value in each individual case

and because of the wide range of normals, it is well to keep in mind that protrusion on one side is a possibility. As mentioned previously, appearances are deceiving in the thyroid type of exophthalmos and great care should be taken in measuring and recording the progress of the protrusion.

Decrease in size of the orbit: The orbit reaches its adult size and shape fairly early in life, and although it expands slowly with normal growth of the individual, the relative proportions of the orbit to its contents remain the same. Therefore, any condition that diminishes the size of the bony vault pushes the contents out from the base of the cone and produces exophthalmos. Roentgen examination is very important in this group of cases because changes within the walls of the orbit can be visualized and comparative measurements can be worked out.

A common cause of decrease in the size of the orbit is trauma. Compression produces displacement of the orbit and a greater displacement of its contents. The eye may be very prominent for a time but slowly the orbital tissue atrophies and later there is a pronounced enophthalmos on the injured side. Other types of injury may bring the eye forward but, in most of these cases, atrophy and enophthalmos result. When the orbit and globe are displaced, a problem in the fitting of glasses arises which must be considered carefully. This is in the adjustment for the "off position eye." The patient may require a vertical prism and a decentration of the lens and a dropping of the frame on one side.

Changes in the bony vault are not very common causes of exophthalmos although we have seen cases of *hyperostosis* which involve the greater wing of the sphenoid and push the eye forward. Roentgen examination clearly defined the part of the vault which was involved, but no cause could be found, and the use of deep roentgenotherapy did not change the picture. Exostosis and sclerosing osteitis also may decrease the size of the orbit.

Endotheliomata of the vault of the skull sometimes produce an exostosis and displace the eye forward and downward.

Paget's disease may cause exophthalmos and loss of vision at the same time by involvement of the nerve in the canal. This condition is rarely seen, however, and is mentioned only as a possibility.

Bony growths *invading from the cranial vault* decrease the size of the vault and increase the content as well. Here again, roentgen examination will show the existing pathology in most instances.

Exophthalmos is a relatively common eye sign and it is always a sign of a disease process, either at the present time or at some past time. The diagnosis in many instances is not easy to make and the aid of

most of the other fields of medicine may be required. The notes presented here are mostly the result of my own observations and difficulties in diagnosing and treating this serious condition. The problem is to correct the primary cause and obtain a good cosmetic result with a functioning globe. The effort is worth the trial but most of the effort should be expended early—the earlier the diagnosis, the earlier the treatment which results in less protrusion, less residual change and the best functional result.

Patients with a protruding eye or eyes are never satisfied even though the primary condition is arrested or cured. If these notes prevent a single patient from the disfiguration and loss of vision due to exophthalmos, they have served their purpose.