the formation of the Committee whose report submitted in October, 1941, constituted the plan of the work that was performed subsequently.

Special reference should be made here to the kind support to this work of H.E. Lady Jacqueline Lampson (Killearn).

For the details of the work achieved, the archives of P.H.M., Cairo, should be consulted. Only a brief outline is given in this paper, read at the meeting of the Ophthalmological Society of Egypt.

It is to be hoped that this traditional collaboration of efforts in the fight against trachoma will be continued not only in Egypt but also outside this country, on a wider scale, now that the war is over and bigger fields of anti-trachoma work will soon be opening up.

PROPTOSIS—DIFFERENTIAL DIAGNOSIS

BY

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DIAGNOSIS of cases of proptosis is of importance to the physician and surgeon as well as to the rhinologist and ophthalmologist. In fact, here is a problem which touches different fields of medicine and for which co-ordinate working of men of different specialities is often necessary.

By the term proptosis is meant a state of protrusion of the globe of the eye between the lids. Exophthalmos is synonymous. Beer suggested calling the affection exophthalmus when the protruded eye is in its natural state, exophthalmia when it is inflamed and exophthalmoptosis when the displacement is caused by division of the nerves and muscles of the orbit or by paralysis of the latter.

As is well known an end diagnosis of a case of proptosis is not always easy. For this reason, it will be found helpful if as a first step an attempt is made to place the case to be diagnosed in one of six main types or categories. These are:—1. Apparent; 2. Due to a congenital anomaly; 3. Due to a traumatic injury; 4. Due to an inflammatory process; 5. Due to a new growth; 6. Due to systemic trouble or disease.

It will not be difficult usually to place a case, from the general examination and the history, in one of these main types, this notwithstanding the possibility of an overlapping, as in cases that are due to a trauma and inflammation together, or in cases that present a congenital origin with a new growth, etc. And so having

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placed our case in the most likely category, we proceed to differentiate between the states of the one type under consideration. For this purpose it will be found necessary to use different methods of examination. The nearest are the ophthalmological and rhinological, and then the X-ray appearances, blood tests and blood cytology. The remotest include special laboratory studies, medical and surgical procedures.

Under type I.—the apparent cases of proptosis, the globe is not really displaced. There are the cases of high myopia, buphthalmos, megalophthalmos and staphylomata¹. Also cases of 7th nerve paralysis where the palpebral fissure gets wider with retraction of the lids.

In type II.—that which has a congenital anomaly, we have those cases of imperfect development of the orbit, cases of premature ossification, synostosis of coronary or sagittal sutures in oxycephaly, whereby the growth and increase in size of the brain, pressure is exerted on the orbit from the back with consequent proptosis of different degrees up to a complete forward dislocation². This may be associated with divergence of the eyes, pressure atrophy of optic nerve or a choked disc. When arrested development occurs along the lines of the orbito-nasal fissure, fronto-ethmoidal or sphenoidal sutures, thus leaving a defect in the orbital wall, then a hernia is the result; meningocele, encephalocele, encephalocystocele, coming into the orbit with consequent proptosis. All these appear as tumour masses at the upper and inner angle of the orbit, reducible if the aperture in dura is not closed.

Type III.—With cavernous sinus thrombophlebitis, swelling of lids is followed soon by exophthalmoplegia, 3rd, 4th and 6th nerves passing through the sinus are usually affected. There is marked distension of retinal veins; and the condition becomes bi-lateral very soon. The sinus may be affected by way of petrosal sinuses in middle ear disease¹¹. Exophthalmos may then be moderate and transient as venous congestion subsides. When infection is found in the ear region, exophthalmos with chemosis may be elicited by pressing on the neck with a sand-bag if the cavernous sinus is involved.

Type IV.—Inflammation of nasal sinuses frequently involves the orbit, especially with ethmoid cells which are separated from the orbit by the lamina papyracea¹², with consequent orbital cellulitis, sub-periosteal abscess, orbital or retrobulbar phlegmon.

With a state of subacute or chronic inflammation of the sinuses, a mucocele is formed and this is a frequent cause of unilateral proptosis¹³. Congestion occurs in the orbital cavity on the affected side with chemosis of conjunctiva, redness of lids, headache and fever. Vision is reduced, globe is fixed and proptosed in a direction away from the site of disease¹⁴. In ethmoid disease it is pushed laterally, in frontal sinus affections it is pushed down with tenderness

at the orbital margin. Choked disc is usually seen and panophthalmitis may follow. The onset of proptosis is gradual in a case of mucocele until it ruptures when it may produce sudden exophthalmos.

Other inflammatory conditions with proptosis are orbital periostitis (pneumococcal) which is more frequent and less serious than the tuberculous form with sequestra, fistulae and possibly tuberculides of the skin. Both are to be diagnosed from syphilitic periostitis with early bone hyperplasia, and gumma with deep seated pains.

Under proptosis with inflammatory conditions come also those cases in which it is noted to occur after infection has passed and is due to impeded circulation in the orbit. Also those known as pseudo-tumours from low-grade non-mycotic inflammation, as well as cases of strepto-actino-mycosis with fistulae, anthrax, hydatid cysts, and trichinosis with orbital muscle pains—all of which need special laboratory studies for diagnosis.

Type V.—Cases of New growth—these include:—

a. Tumours of intracranial origin which produce exophthalmos when pressing on a part of the orbit or subsequently penetrating into its interior. Unilateral proptosis has been observed to occur in about 2 per cent. of all cerebral tumours; uni- and bilateral in 8-10 per cent. of cases. The most frequent that cause exophthalmos are tumours of the base of the skull, frontal and temporal lobes, *i.e.*, those that have a direct connection with the orbit or that can determine increased tension in the middle and anterior cranial fossae. A meningioma, in Cushing's syndrome with orbital hyperplasia and thickening of the posterior wall, takes a slow course with no fundus changes. It is the cause in about 50 per cent. of unilateral¹⁵ proptosis, then orbito-ethmoidal osteoma, then frontal bone osteoma.

Pituitary tumours eroding the sella and penetrating into the orbit cause proptosis with symptoms peculiar to that locality, hemianopia, acromegaly, etc. Tumours of the cribriform plate between the two orbits produce exophthalmos with mental signs resembling those of frontal lobe tumours; symptoms of a retrobulbar neuritis, central scotoma, with diminished vision may occur.

Tumours in and about the sphenoid fissure¹⁶ compress motor and sensory nerves there, giving rise to ptosis, divergent squint, mydriasis and diminished cutaneous sensations in the face. Failing vision met with in these cases is usually long delayed until pressure is exerted on the optic nerve in or about the optic foramen.

b. Tumours from the mouth, cystic tumours from paradental epithelial debris and from post-nasal cavities, in the nasopharynx and nasal accessory sinuses, myxomata, chondromata, adenomyxomata, plasmomata, all cause proptosis when they extend to the posterior part of orbit through neighbouring structures, antrum or

ethmoid cells. With those that fill the posterior nares the patient has a peculiar look with mouth breathing and nasal intonations. New growth from nasal accessory sinuses, osteomata from antrum and frontal sinus reaching orbit by extension produce exophthalmos very gradually. The direction of displacement of the globe with extent of motility, transillumination and X-ray appearance of the orbit and surrounding structures help in locating the site of a tumour.

c. New growths originating from the orbit include carcinoma from the lacrimal gland growing between periorbita and bony wall; also cylindroma occurring as part of parotid mixed tumour in the lacrimal gland region, with absence of pain and histological structure resembling salivary tumour. Osteomata and fibromata from bone and periosteum, hard, resisting to backward pressure on globe, in contrast to a cavernous angioma which is soft and easy to press.

d. Ocular new growth. In primary tumours of optic nerve and sheath, proptosis occurs in about 7 per cent. of cases preceded usually by failing vision and white atrophied papilla. New growth occurring as such in the muscle cone pushes the globe directly to the front. Vision may remain unaffected if the optic nerve is not involved, as in vascular new growth with free play of muscles and not much pain. With those coming from inside the globe as choroid tumours, the fundus cannot be illuminated, vision is lost and pain is present. A complicated cataract is common. Glioma tumours occur in young children, showing peculiar fundus appearance and blindness early in the disease.

e. Metastatic new growth—sarcoma and carcinoma have been mentioned as causing proptosis with malignant disease of the kidney in children¹⁷. Of great help in diagnosis of all cases of new growth is to note in X-ray pictures the presence of orbital dilatation and its significance whether primary or secondary, also the faint shadows of tumour masses, with marks of pressure atrophy on adjacent tissues and of erosion when the growth is malignant.

VI. Proptosis connected with systemic troubles has an intimate relation with endocrine activities and with the sympathetic nervous system. The mechanism of action is through venous dilatation associated with nervous stimulation of the orbital contents. It is noted to appear as a transient protrusion of the eyes in sudden and violent emotions, in cases of angio-neurotic oedema, during menstruation and in child-birth. Such temporary protrusion of the globe could be induced by ephedrine which is a specific sympathetic excitant and inhibited by yohimbine, a depressant to the sympathetic.

Transient proptosis may be the first sign of a threatening exophthalmic goitre. This disease, known also as Basedow's and

Graves' disease, though connected with the thyroid, is believed to be not a primary disease of that gland. Increased thyroid secretion facilitates the appearance of exophthalmos and vice versa, hypothyroidism induces enophthalmos¹⁸. For the diagnosis of Graves' disease there are the classical signs, von Graefe's, Stellwag's, Moebius', Gifford's and Dalrymple's. Fine tremors of the extended hands occur, with nervousness and increased pulse rate. Exophthalmos may be associated with oedema of the conjunctiva. The orbital contents show vascular dilatation, cellular invasion in the perivascular lymph spaces, oedema of tissues and sometimes increase in orbital fat with some hypertrophy of ocular muscles. The condition may be uni- or bilateral.

Of congenital origin also are some vascular tumours of the orbit, lymphangiomas, dermoid and retention cysts in which proptosis may be associated with optic atrophy³. They occur near the lacrimal gland region and are usually compressible when palpable⁴. Their onset at an early age, with slow development if any, absence of other pathological signs together with X-ray pictures help in their diagnosis.

Of tumours that have a congenital background or predisposition are the neurofibromata or plexiform neuromata that grow by proliferation of peripheral nerve endings⁵. They produce exophthalmoptosis with possibly ocular motor disturbances.

Of systemic diseases that have a congenital origin and in which proptosis may make its appearance is van der Hoeve's syndrome of blue sclerotics. The pathology is said to be endocrine with a disturbance of calcium and phosphorous metabolism⁶.

III. Traumatic cases include wounds of the orbit with possibly foreign bodies retained there, fractures of the base of the skull, injury to blood vessels and to the cavernous sinus, also injuries during birth in infants. With a history of injury, the classical signs may not appear all at once, there may be crepitations, emphysema, lacerations of extra-ocular muscles, orbital haemorrhage, and impaired ocular motility. Pulsating exophthalmos makes its appearance in such cases. The lesion may be intracranial, connection taking place between the internal carotid and the cavernous sinus—intra-orbital with aneurysm of the ophthalmic artery—or extra-orbital with connection between carotid and jugular. It may be possible to observe modification of bruit, exophthalmos and pulsations when temporary pressure is applied over the carotid in the neck.

In rupture of the carotid into the cavernous sinus, the first symptom is usually abducens paralysis, then unilateral exophthalmos⁷. There may be no pulsation but only a swishing sound over the temples. Later fundus changes occur with dilatations, tortuosity of veins and increased intra-ocular tension.

Proptosis when unilateral, pulsating, progressive and reducible

is almost always due to arteriovenous aneurysm; when intermittent, nonpulsating, it is orbital varicocele³ that follows position of the head. It has been noted to be nine times more frequent in the left orbit than in the right. Vision is not affected except rarely when complications occur or haemorrhage.

After trauma, thickening of the bones of the orbit underneath the periosteum may occur with proptosis, from encroachment on the globe. It is met with in the frontal, maxillary or sphenoidal bones. The growth is slow and vision is not much affected. There is proliferation and hypertrophy with no material change in the structure of bone.

IV. Proptosis with inflammatory conditions may follow injuries or occur *per se*. With acute inflammation, conditions are met with, varying from a simple dacryo-adenitis to penetrating wounds of orbit associated with septicaemia, septic thrombosis of the cavernous sinus and lateral sinus and tetanus. Infection may occur through the nose, the pharynx, the nasal sinuses or other inflamed neighbouring structures around the orbit. There are then the signs of infection—pain—often severe, redness, oedema of lids, chemosis with the upper lid falling down on the proptosis, also raised temperature and toxæmia.

An orbital abscess may form remaining unilateral, fixing the globe or rendering its movement painful⁹. Acute orbital cellulitis may begin on one side and rapidly extend to the other or toward the brain with consequent raised intracranial tension, inadequate venous drainage, engorgement and oedema of orbital tissues and proptosis¹⁰. Chemosis of conjunctiva with swelling of lids, 6th n. paralysis, reduced vision and toxæmia are usual signs. A cytological blood count is of help.

In ordinary cases of Graves' disease, exophthalmos may persist in spite of medicinal and surgical treatment; the palpebral fissure is enlarged with frequently inadequate protection of the eyeball.

Malignant exophthalmos¹⁹ is a serious condition occurring with thyroid pathology when the basal metabolism is within normal limits. It is supposed to be due in part to pituitary activity or in fact to a general polyglandular upset.

With pituitary pathology uni- and bilateral exophthalmos has been described, associated with retrobulbar venous stasis, a condition being most marked with the eosinophilic form of oedema of pituitary²⁰.

With vitamin C deficiency and infantile scurvy, haemorrhages in the orbit²¹ producing exophthalmos occur. Proptosis appears suddenly and may be very pronounced. Also in rickets and haemophilia where the subperiosteal haemorrhages may be associated with extravasations in the skin of the lids.

In leukaemias and lymphoblastic allied conditions, infiltrations

with lymphocytes, endotheloid and giant cells occur²², also in Mikulicz's disease with lacrimal and salivary gland enlargement. In all these cases a blood cytology is needed, for diagnosis.

Other hyperplastic processes invading the orbit and producing exophthalmos occur in the rare conditions of xanthomatosis, Schüller-Christian-Hans disease in young children. Cholesterin deposits in various organs and in skull bones occur with degeneration, diabetes insipidus and cessation of growth. Pressure and loss of bony support at the apex of the orbit produce proptosis usually bilateral with fundus changes and loss of sight.

In Paget's disease, exophthalmos occurs when bony enlargement is well advanced, also in leontiasis ossea occurring earlier in life and more often in females.

Cases

1. A. S.—boy aged 15 years—came with bulging of L. globe that showed as proptosis when the lids closed. It was only a staphyloma that extended over almost the whole cornea. An iridectomy was performed with puncture of the head of the staphyloma and a pressure bandage applied. The globe returned to a normal level within a week.

2. M. O.—boy aged 15 years—came complaining of a foreign body, a speck of dust stuck on the cornea. He presented a noticeable bulging of both globes dating from childhood. Neither his vision nor the globe movements were much affected. This case was of interest as an example of a congenital shallow orbit which was evident on X-ray examination.

3. F. O.—man aged 25 years—had a gunshot wound in the right temple. Sixteen hours later, he presented right bulging of the globe which was increasing with oedema of the lids and chemosis all extending to the other side and accompanied with toxæmia. The diagnosis of cavernous sinus thrombosis was confirmed by operation.

4. A. M.—boy aged 5 years—came with proptosis of the right globe that began three days before, first slight then rapidly increasing. Lids of right eye red and swollen, globe projecting to the right, conjunctiva swollen and congested with restricted and painful movements of the globe. Fundus normal and vision not affected. History of recurrent nasal catarrh. Slight fever and enlarged and tender pre-auricular glands on the same side. All the signs denoted the cause as being an orbital abscess. Diagnosis confirmed by exploratory puncture, and evacuation of pus followed by recovery.

5. F. S.—woman aged 40 years. Left proptosis down and out with limited movements up and out. Vision not much affected. Fundus appearance normal except for somewhat dilated veins. A

cystic swelling could be felt around the upper part of the orbit. The skin of lids normal, with no distended veins, no roughness of bony edges of orbit, no pulsations and not much pain. Rhinological and X-ray examination suggested mucocele of the left frontal sinus with absorption of the orbital wall. Diagnosis confirmed by operation.

6. B. H.—woman aged 50 years—came with the complaint of severe headache of two weeks' duration, more on the right side. Right globe proptosed with a slight restriction of movement, and failing of vision. History of vomiting unrelated to food. Fundus showed dilated veins and oedema of the papilla. Rhinological and X-ray examination showed nothing in particular. W.R. strongly positive. Diagnosis, probably gumma. It was confirmed by improvement on antisyphilitic treatment.

7. R. A.—a girl aged 13 years—presented a proptosed left eye with no vision. Fundus opaque, severe headache and vomiting. Diagnosis in this case was helped by the history of enucleation for glioma in the other eye six years before. Patient died one month later.

8. A. F.—a woman aged 35 years—presented proptosis of left globe. A mass could be felt at the upper and outer part of the orbit in the region of the lacrimal gland, vision not much affected. There was a diffuse swelling in the region of both parotid glands. Blood cytology showed increased lymphocytes. W.R. was positive, but no improvement on antisyphilitic treatment. This case was probably of the type of Mikulicz's syndrome.

Summary

1. Apparent—globe not displaced; high myopia; buphthalmos; megalophthalmos; staphyloma; 7th nerve paralysis.

2. Due to congenital anomaly—arrested development; cranial hernia; premature ossification; shallow orbit; tumour masses; vascular, nervous, mixed.

3. Due to traumatic injury—intracranial, tear of internal carotid and cavernous sinus; intra-orbital, aneurysm of ophthalmic artery; extra-orbital, tear of carotid and jugular; late effect, proliferation.

4. Due to inflammatory process—acute, from injury or extension, abscess, cellulitis, thrombophlebitis, anthrax; subacute, mucocele, retrobulbar, subperiosteal; chronic, tubercular, syphilitic, actinomycotic, hyatid, etc.; late effect, from obstruction.

5. Due to new growth—intracranial—meningioma, osteoma; oral, nasopharyngeal—myxoma, chondroma, plasmoma, osteoma; orbital—cavernous angioma, osteoma, lacrimal cylindroma, carcinoma; ocular—from optic nerve and sheath, choroid, retina; metastatic—sarcoma, carcinoma.

6. Due to systemic condition—sympathetic—Graves' disease; endocrinal, malignant exophthalmos; metabolic—scurvy, rickets, haemophilia; hyperplastic—leukaemias, xanthomatosis, Paget's disease.

Methods of diagnosis—general exam. history; transillumination; X-ray pictures; blood tests, cytology; special laboratory studies; medical and surgical procedures.

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ANNOTATIONS

Dissatisfied Patients

One of the great problems of medical practice is what to do with our dissatisfied patients. Happy is the practitioner who can truthfully say he never had any. Years ago at a Moorfields' Students dinner Sir Anderson Critchett was in the chair. His health was proposed by Mr. William Lang who said, among other things, that he always wondered what Sir Anderson did with his dissatisfied patients, as he never saw any of them. Sir Anderson refused to be drawn by this blandishment and the juvenile ophthalmologist got no useful tips as to how to deal with this troublesome question. One supposes that each of us in time and with sufficient practice evolves our own methods of dealing with dissatisfied patients. Much depends on the cause of dissatisfaction. Sometimes it is mutual, and then the best