central retina was considered at one time to be the result simply of a spread of the disease from the peripheral region. But sporadic reports have appeared which indicate a primary lesion of the central retinal vein within the optic nerve-head, and this central retinal vasculitis was described in detail by T. K. Lyle and K. Wybar.[3] The condition, which is usually unilateral, shows several characteristic features: a considerable degree of oedema of the optic disc, which may extend into the macular area; a massive dilatation of the retinal veins extending from the optic disc to the periphery; a variable amount of retinal haemorrhage, sometimes with the appearance of peripapillary cotton wool spots; an almost invariable absence of vitreous haemorrhage; and remarkably little disturbance of the vision (early or late) except when the macular area is involved in haemorrhage or, when the retinopathy is severe, by a form of cystic maculopathy. Eventually there may be some sheathing of the retinal veins, but this is variable, and the only conspicuous late features are the indefinite persistence of some degree of dilatation of the retinal veins, a post-oedematous blurring of the optic disc, and occasionally some disturbance of the macular area.

In a recent comprehensive study of central retinal vasculitis C. D. Hart and colleagues found evidence of a normal retinal arterial pressure (measured by ophthalmodynamometry) and by normal arterial filling (studied by fluorescein angiography), except in one of the nine cases in which there was an associated arterial disease due to malignant hypertension. But there was an obvious abnormality of the retinal veins as shown by the absence of a spontaneous retinal venous pulsation in the affected eye in all cases, by a grossly raised retinal venous pressure in five of the nine cases, and by evidence of a vascular decompensation, which assumed two main patterns. Firstly, a stage of venous decompensation was noted, with dilatation of the retinal veins and leakage of fluorescein dye from the larger veins (perivenous fluorescence). At this stage the retinopathy was mild or moderate—retinal haemorrhages, cotton wool spots, minimal retinal oedema, and more or less normal vision. Secondly, a stage of capillary decompensation, with dilatation of some of the retinal capillaries, often with the formation of microaneurysms, and with extensive leakage of the dye particularly in the macular area. At this stage the retinopathy was severe—considerable retinal oedema and changes in the macular area which resembled those of a cystic maculopathy in addition to the other features of a mild or moderate retinopathy. There were also areas of capillary closure and the appearance of shunt vessels at the optic disc.

It is evident that a reduction in the arteriovenous pressure gradient (perfusion pressure), which is produced solely by a rise in the venous pressure, is a characteristic feature of central retinal vasculitis. It results in a rise in tissue pressure in the retina, which accounts for the areas of capillary closure. It probably also causes an ischaemic cellular oedema and a failure of the tissue fluid to be absorbed by the venous capillaries, since the intraluminal pressure exceeds the plasma protein osmotic pressure. The severity of the retinopathy is almost certainly determined by the site of the lesion in the central retinal vein. When the lesion is proximal to the optic disc the retinopathy is mild—that is, it assumes the form of a venous decompensation—but when it is within the optic nerve or optic nerve-head the retinopathy is much more severe—that is, it assumes the form of a capillary decompensation.

Hart and his colleagues suggest that central retinal vasculitis represents a form of central retinal vein thrombosis in association with an otherwise normal vascular system, so that there is a potential for the development of collateral channels. Hence the retinal changes are less severe and the visual prognosis is better than in the usual forms of central retinal vein thrombosis. This is an interesting concept, and thrombosis almost certainly is an element of some of the more severe cases. But certain points cast doubt on such an emphatic view. An example is the occurrence of central retinal vasculitis in one eye and of peripheral retinal vasculitis (Eales's disease), which is certainly not the result of a thrombotic state, in the other eye. Again, a massive increase in the retinal haemorrhages may follow the use of anticoagulants, yet they may respond favourably in many cases to systemic steroids.[3] A response to steroids was not experienced by Hart and colleagues,[4] who favour the use of acetazolamide in an attempt to improve the ocular perfusion.

Certainly the differential diagnosis of central retinal vasculitis is more or less limited to central retinal vein thrombosis, if it is accepted that the two conditions are not identical. As a general rule the retinal haemorrhages in a central retinal vasculitis are less than would be expected in a central retinal vein thrombosis in view of the extent of the disc oedema and the massive retinal vein dilatation which are features of both conditions. Other conditions may be mentioned but they should not cause confusion: plerocephalic oedema is bilateral (as a general rule) and the dilatation of the retinal veins is confined to an area round the optic disc; papillitis is characterized by a distinct loss of vision in the acute stage; and a fulminating hypertensive retinopathy is bilateral and shows other obvious features of widespread arterial disease.

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Food and Cyanide

In some regions of the world diseases which are relatively uncommon or unknown elsewhere are frequently encountered. There may be many different causes for this, including racial and genetic factors as well as infective agents. If all of these can be safely excluded, attention naturally turns to the food and drink. At times the food incriminated may be esoteric in the extreme. The eating of close relatives' brains in New Guinea, for example, led to the transmission of the fatal disease of kuru. At other times the food may be commonplace, as in some recently investigated tropical neuropathies.

For many years an ataxic neuropathy has been observed in the tropics unlike that due to the common vitamin deficiencies. The patients have a loss of joint position sense leading to a sensory ataxia. This has been ascribed to a posterior column myelopathy, though few necropsies are available.[1 2] There is a relatively slight loss of superficial sensation or impairment of lower motor neurons. The sphincters are only rarely affected, and it is uncommon for there to be much impairment of upper motor neurons. The

lower limbs are particularly involved, and the condition is slowly progressive. Bilateral nerve deafness often preceded by tinnitus, and bilateral visual deterioration leading to optic atrophy, are both common in this condition. It has been reported that the peripheral nerves show segmental demyelination,[3] though some further evidence for that may be desirable.

With the interest in cyanide metabolism arising from studies of tobacco amblyopia and Leber's optic atrophy[4-6] attention turned to the possibility of chronic cyanide intoxication as a possible cause of this condition. It was immediately recognized that the staple diet in many endemic tropical areas was a root or other vegetable rich in cyanogenic glycosides. Thiocyanate is found in high concentration in the blood of patients with this disease, and it has been suggested that this arises as a result of detoxification of cyanide.[1 7] A recent study of a similar condition in East Africa has confirmed the findings in Nigeria.[8] In the latter country it has been shown that patients with the neuropathy have a higher intake of cassava than people who did not develop the neuropathy.[1 7] The clinical improvement resulting from a reduction of the amount of cassava in the diet and the substitution of non-cyanogenic foods confirms this association.

The exact role played by vitamin B_{12} in the neuropathy is not clear, despite considerable discussion. The total serum vitamin B_{12} level is high in patients with a toxic tropical neuropathy.[1 8 9] The proportion of vitamin B_{12} present as free hydroxycobalamin and as cyanocobalamin still requires to be defined. Though it has been suggested that vitamin B_{12} has a primary role in the detoxification of cyanide, this is not finally proved. Much remains to be elucidated about the effect of cyanide on the nervous system. For instance, an entirely different symptom complex of amyotrophic lateral sclerosis, Parkinson's disease, and dementia is seen in Guam.[10] It has been suggested that this results from intoxication with cycasin, a cyanogenic glycoside present in food prepared from the cycad nut. Experimentally this glycoside causes a cerebellar degeneration in experimental animals.[11]

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Duodenal Diaphragm

The commonest cause of obstruction to the gastric outlet in the adult patient is peptic ulceration, usually duodenal, but occasionally in the pyloric or prepyloric region of the stomach. Second to this comes carcinoma of the gastric antral region. Other obstructive lesions are numerous but rare. They include adult pyloric hypertrophy, benign tumours, infiltration by one of the reticuloses or an adjacent carcinoma of the pancreas, heterotopic pancreatic tissue, and prepyloric mucosal diaphragm.[1] One of the least common but nevertheless interesting causes is a persistent congenital diaphragm in the second part of the duodenum.

Congenital duodenal occlusion presenting as a neonatal emergency may be due either to a diaphragm or to a complete obliteration of the duodenal lumen, which ends as a cul-de-sac. Though the latter may result from vascular occlusion in utero, it is probable that the diaphragm represents incomplete vacuolation of the proliferating epithelial lining of the duodenum, which leads to its temporary obliteration in the sixth and seventh weeks of fetal life.[2] About 60% of cases have some other congenital anomaly, and about one-third of the patients have Down's syndrome (mongolism).

If the occlusion is complete, symptoms appear within hours of birth. But in some cases a lumen may be present in the diaphragm, so that life is sustained for months or even years. Rarely the lumen is wide enough to allow survival into adult life. Long-standing incomplete obstruction may result in dilatation of the proximal duodenum,[3] and in some cases the mechanical action of the food may balloon the septum into the distal duodenum like a windsock to produce an intraluminal duodenal diverticulum. Zatzkin and his colleagues[4] have given a clear description of the radiological appearances of such a case (the third to be reported), and a remarkable example of such a diverticulum was reported by A. J. Levi and L. Kreel[5] in a young man of 24 who had undergone gastrojejunostomy at the age of 6 days for what was thought to be a duodenal atresia but which, in adult life, proved to be a duodenal diaphragm.

J. L. Bilton and S. Yap[6] have recently summarized the features of duodenal diaphragm as well as reporting two more examples of this condition. The history, in adults, is usually one of persistent vomiting. The vomitus may or may not contain bile, this depending on whether the diaphragm is below or above the ampulla of Vater. There may be epigastric distention and loss of weight, and there may be melaena or haematemesis. Though the diagnosis is not often made preoperatively, careful *x*-ray studies, with hypotonic duodenography, may show the obstruction of the second portion of the duodenum with a small trickle of contrast material passing through the orifice in the diaphragm.

In a condition as rare as this it is not possible to be dogmatic about surgical treatment. Though duodenojejunostomy is regarded as the treatment of choice in infants, F. D. Threadgill and A. Hagelstein[3] report two cases in which this procedure was done and met with failure. One patient was left without relief, the other failed to respond to subsequent gastrojejunostomy and was eventually cured by partial gastrectomy. C. N. Hudson[7] reported a case in which excision of the diaphragm and transverse closure of the duodenum was followed by excellent results, and others have had similar experiences.[4 5] Of the two patients reported by Bilton and Yap the first responded well to this simple excision with duodenoplasty, but the second, suffering from severe gastrointestinal haemorrhage, was treated by vagotomy and gastrojejunostomy, with equally good results.

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