Not all cases of retinitis pigmentosa are hereditary, and there are many atypical forms, including those where the clinical changes are limited to one eye, one sector of the fundus, or present as a "retinitis pigmentosa sine pigmento." It has been suggested that this disease may really be little more than a syndrome, and that damage to the visual cells may be provoked by quite disparate causes, since the specialized tissue of the retina can react in only a limited way; and, furthermore, since the antigenicity of the retinal photoreceptors has been generally established, an immunological injury might well be responsible for such degenerative changes.

There have been several recent studies of the biochemical features of retinitis pigmentosa, which generally indicate an increase in the immunoglobulins,[2] especially IgM, suggesting an infective or autoallergic process. The histological appearances rarely show any evidence of inflammatory reaction, but the activity of slow viruses has been known to provoke macular damage, and such an occult virus infection might well underlie some cases. Rahi[2] concluded in a recent survey that the raised IgM level might either cause the retinal degeneration directly or through the intermediary of such an occult (? slow virus) infection, or that individuals with a raised IgM (whose production is genetically controlled) might be more prone to retinal degeneration, or else that the high IgM level might simply represent a further expression of the retinitis pigmentosa genetic anomaly.

[1] Jay, B., and Bird, A., *Transactions of the American Academy of Ophthalmology and Otolaryngology*, 1973, 77, 641.
[2] Rahi, A. H S., *British Journal of Ophthalmology*, 1973, 57, 904.

Cold and the Heart

The atmospheric climate of Britain, if not the financial, appears to be improving. Lulled by two relatively mild English winters it may be salutory to remind ourselves of some of the hardships of our cardiac patients in prolonged periods of cold weather. Most doctors are familiar with the problems that a patient's angina pectoris may become worse in cold weather, and stepping out into the cold air first thing on a winter morning may precipitate a sharp attack of chest pain on minimal exercise. Physicians who see large numbers of hypertensive patients have long had the impression that the blood pressure of their patients tends to be higher on cold winter days than in the balmy days of June, and this has recently been confirmed.[1] The two observations have been linked in a recent report from Portland, Oregon, which suggests that there may be a correlation between exacerbations of angina pectoris and this rise of blood pressure produced by exposure to cold.[2]

Neill *et al.*[2] set out to find the mechanism of exacerbation of angina by the cold. In an experiment there are many ways of applying a cold stimulus—locally, diffusely to the skin, or by the inhalation of cold air; but the haemodynamic responses appear to be similar in each instance.[3] The method chosen in this instance was to place, for either two or six minutes, a damp towel filled with ice chips on the foreheads of 19 patients with coronary heart disease and 6 control patients, all of whom were undergoing left heart catheterization for diagnostic purposes. All consented to the experiments with cold as a separate risk. In the patients exposed to cold for six minutes local discomfort and pain was maximal between one and two minutes of exposure and subsequently decreased. Five patients with ischaemic heart disease (all of whom gave a history of cold intolerance) developed typical angina pectoris after two to three minutes exposure. Parallel with the local discomfort there was a marked increase in both systolic and diastolic blood pressure and a slight increase in heart rate, which was maximal after some six minutes exposure. Coronary blood flow, measured by a xenon disappearance technique, was also increased by the cold and was greater at two minutes than six minutes. As well as changes in systemic arterial pressure cold exposure produced increases in left ventricular end diastolic pressure and left ventricular myocardial oxygen consumption (calculated as the product of coronary blood flow and coronary arteriovenous oxygen difference). All these changes occurred in both groups of patients studied—controls and those with ischaemic heart disease. In seven of the latter exposure to cold resulted in evidence of myocardial hypoxia—an increase in the lactate/pyruvate ratio in coronary venous blood of more than 15% over the resting level.[4] These changes suggestive of myocardial hypoxia were not, however, confined to patients with a history of cold intolerance.

A different form of stress to the myocardial oxygen supply —production of a tachycardia by atrial pacing—produced the same evidence of myocardial hypoxia as did cold exposure in the patients with ischaemic heart disease, suggesting that their myocardial hypoxia seems not to be a special response to cold but simply manifestation of diminished coronary reserve.

So it seems that the increase in systemic arterial blood pressure produced by exposure to cold causes an increase in oxygen consumption by the heart. In patients whose coronary circulation is already compromized this extra demand produces myocardial hypoxia and angina pectoris results. This explanation has a better physiological ring to it than previous hypotheses such as "coronary artery spasm," for which there is little factual evidence.

What advice can we give patients with angina pectoris produced by cold exposure? Apart from the rather impractical suggestion of moving to warmer climes, they should be reminded to take their glyceryl trinitrite before stepping out on a cold morning. Further, since β-blocking drugs help the patient with angina by diminishing myocardial oxygen requirements a seasonal increase in the drug dose might be appropriate.

[1] Dollery, C. T., *British Medical Bulletin*, 1973, **29**, 158.
[2] Neill, W. A., *et al.*, *American Journal of Medicine*, 1974, **56**, 471.
[3] Leon, D. F., Amidi, M., and Leonard, J. J., *American Journal of Cardiology*, 1970, **26**, 38.
[4] Neill, W. A., *American Journal of Cardiology*, 1968, **22**, 507.

Squint

A squint simply means a deviation of the visual axes (not, as people often imagine, eyes that look obliquely, are half-closed, or inspect at a very close range); but not only is the name confusing—hence the occasional use of that clumsy classicism strabismus—but the subject is so complex that international congresses and frequent symposia have become regular features of the ophthalmological scene.

Such deviations are usually in the horizontal plane and may be convergent or divergent. They may be obvious if

Br Med J: first published as 10.1136/bmj.3.5928.430-a on 17 August 1974. Downloaded from http://www.bmj.com/ on 19 April 2024 by guest. Protected by copyright.

gross, but small degrees of deviation can be recognized from the asymmetrical positions of the bright corneal light-reflections relative to their respective pupil margins; they can then be confirmed by covering each eye in turn and noting whether a perceptible movement is made by either eye to assume fixation when its fellow-eye is occluded.

The eyes are kept parallel by a complicated conditioned reflex acquired during the first few years of life in the interests of single vision; this reflex is consolidated when the child learns to fuse the two superimposed images from either eye and finally to appreciate stereoscopic vision. The squint is labelled paralytic if the motor-apparatus that rotates the eyes is damaged. It is labelled concomitant if there is an impediment to the sensory component of the reflex arc (such as a poor-sighted eye from any cause) or to the central component of the reflex arc (such as mental deficiency or a psychic disturbance).

The management of most paralytic squints is straightforward, since they tend to derive in older patients from evident intracranial damage and gradually recover when the latter is rectified. They rarely require more than symptomatic relief of the attendant diplopia; this can be provided by advice on turning the head or by an occluder on the spectacle lens.

Concomitant squints are more common and more complex; they usually arise in early childhood, and their management is far more important. The first essential is that all cases of suspected squint should be referred promptly for examination by an eye-surgeon. The squint may be the first evidence of other disorders such as a retinoblastoma; it may already be so gross, as in congenital cases, that there is no prospect of the eyes being used in unison, in which case a straightening operation should not long be delayed; it may be illusory (often counterfeited by rather large infantile epicanthic folds), and the parents can then be reassured; and if there is a true squint it is always necessary to prevent the development of amblyopia before it is too late. Amblyopia ("lazy eye") can usually be rectified by occluding the better-seeing eye, and so forcing the child to use the one that squints; this is generally successful before the age of 7 but rarely after the age of 10—hence the urgency of treatment. The underlying tendency to converge should be neutralized by correcting any hypermetropia with appropriate convex spectacles. Infants are very rarely myopic, but in such cases there will be a corresponding tendency to develop a divergent squint. If a marked squint persists in spite of correct glasses and the prevention of amblyopia an operation is usually indicated to re-allign the squinting eye; this normally entails the reinsertion of the medial rectus tendon farther back on the globe—a recession—and an equivalent shortening of the lateral rectus tendon—a resection. Except in congenital cases operation is usually deferred until the age of about 5, by which time the child can also collaborate in orthoptic exercises, which are sensory exercises that help the eyes to build up their fusion and ultimately their stereoscopic vision (and not exercises for the eye-muscles, as the patient usually imagines). Squinting eyes can always be brought straight by an operation at any age for cosmetic effect, but they may well drift out of alignment later if there is a low acuity or poor binocular vision.

A recent symposium[1] at Cambridge has served to clarify many of the outstanding problems, particularly concerning the physiology and psychology of vision, not only in the eye itself but also in the binocular pathways and the cortical organization of visual stimuli. The major damage caused by squints is the production of amblyopia, and the mechanism of this is now far better understood. Indeed, neurophysiological anomalies in the striate and prestriate cortex, and a pathological shrinkage of cells in the lateral geniculate nuclei, have now been shown to occur in monkeys;[2] these striking morphological changes could be caused in one eye if it alone was closed by a tarsorrhaphy, and were reversible if the fellow-eye was in turn occluded. Some amblyopias which develop in apparently normal "straight" eyes have been confirmed[3] as the sequel to a microtropia or microstrabismus in which a deviation of less than 5° is accompanied by a harmonious anomalous retinal correspondence, and, for these, occlusion of the master eye is the essential treatment.

Squints are extremely rare in coloured races and tend to be hereditary and familial. In the past it was thought that the latter reflected poor environmental conditions. In fact,[4] it now seems that social conditions have no bearing on their aetiology; it is just that the children of parents with lower I.Q.s are harder to treat effectively.

[1] Watson, P. G., *British Journal of Ophthalmology*, 1974, **58**, 157.
[2] Noorden, G. K. von, *British Journal of Ophthalmology*, 1974, **58**, 158.
[3] Lang, J., *British Journal of Ophthalmology*, 1974, **58**, 281.
[4] Graham, P. A., *British Journal of Ophthalmology*, 1974, **58**, 224.

Prolonged Intravenous Feeding

From the origin of our species to the past decade the small intestine has been essential to life. Fifteen years ago, for example, after resection of all but 20 cm of the small bowel beyond the ligament of Treitz for superior mesenteric artery occlusion an 80-year-old woman was kept alive for nine months on a high protein, low fat diet, with vitamin and mineral supplements and blood.[1] In spite of all, she was emaciated at death, and necropsy showed intense fatty degeneration of the liver characteristic of starvation. Since then it has become clear that, when not much more than duodenum remains, permanent parenteral feeding is needed, and the necessary techniques have been evolved. In addition to the absolute indication of small-bowel resection there is growing recognition of other conditions—such as severe extensive inflammatory bowel disease, oesophageal stricture, or even severe burns, major trauma, or widespread sepsis—in which prolonged intravenous feeding can make the difference between life and death.[2]

Apart from the practical difficulties of technique[3] several long-term problems have arisen, notably the development of syndromes due to specific nutritional deficiencies not covered by prolonged intravenous feeding with carbohydrate and protein sources of calories, supplemented by the usual vitamins and minerals. The most widely reported of these disorders is characterized by a scaly rash,[4] delayed wound healing,[5] anaemia,[6] increased platelet stickiness[5] (which may be associated with cerebral or coronary occlusion[7]), and a large, fatty liver.[8] This syndrome is due to deficiency of essential fatty acids, and its diagnosis and treatment were recently described in these columns.[7]

In response to the growing need for a safe form of intravenous lipid a 10% emulsion of soya bean oil, Intralipid, containing 56 g/l. linoleic acid was developed, and this provides effective treatment. The body can synthesize arachidonic acid from linoleic acid, which is therefore the only

Br Med J: first published as 10.1136/bmj.3.5928.430-a on 17 August 1974. Downloaded from http://www.bmj.com/ on 19 April 2024 by guest. Protected by copyright.