intravascular infusions of prostaglandin E_2 analogues or combined vasodilator-antithromboxane treatment.

The Raynaud's Association Trust is a most helpful source of information for both patients and their doctors on topics ranging from electrically heated garments and hand warmers to digests of current research. Its publications are available from the secretary (A H Mawdsley, Raynaud's Association Trust, 40 Bladon Crescent, Alsagar, Cheshire ST2 2BG).

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Advances in the treatment of facial deformity

The devastating combination of physical and psychological disability produced by facial deformity is unique. Attempts to correct both congenital and acquired malformations date from prechristian times,[1] but they continue to tax the ingenuity of reconstructive surgeons. Recent advances in technique based on a better understanding of anatomy and physiology have made possible the treatment of some previously inoperable conditions and greatly improved the results in others.

The greatest innovator in this subject since Sir Harold Gillies is Paul Tessier. Stimulated by patients with grotesque deformities associated with severe Crouzon disease he developed radical operations now known as "craniofacial surgery."[2] The two main underlying principles are, firstly, that large portions of the facial skeleton can be mobilised in the subperiosteal plane (that is, devascularised) and moved to a new position and survive; and, secondly, that even when there is gross abnormality of orbital relations (dystopia) the optic foramina are normally related. That means that the walls of the bony orbits may be cut and the eyes repositioned without destroying sight. The manoeuvre requires both intracranial and extracranial approaches and so a neurosurgeon must be brought in.

These techniques are most often used to treat deformities associated with premature fusion of cranial sutures (craniosynostoses). These may be isolated or part of a craniofacial dysostosis syndrome. The surgery is intended primarily to improve the appearance—but its impact is profound. Most patients are of normal intelligence, but because of the severity of their deformities have often been thought to be stupid—with intolerable effects on their daily life from childhood onwards.

Technical advances continue to be made. In particular, operations are being performed at a younger age, 6 months being the optimum for many patients.[3,4] Apart from some practical surgical advantages this approach allows the rapidly expanding brain to be harnessed to mould repositioned bony structures and gives the child the psychological benefits of an improved appearance during his critical formative years. Furthermore, intracranial pressure is sometimes raised in these children without overt clinical signs.[5] Without treatment the raised pressure may be associated with impaired intellectual development, but it is corrected in most cases by early surgery.

The development of a "team" approach implicit in such complex work has produced benefits for patients with many other types of deformity. The combination of plastic, neurological, oral, ear, nose, and throat, and ophthalmic surgeons backed by specialised anaesthetists, radiologists, orthodontists, psychologists, speech therapists, and nurses offers a great breadth of skills. Patients are usually seen jointly for a treatment plan to be established. The parents of a child with, for example, a cleft lip or Treacher Collins syndrome (mandibulofacial dysostosis, which may be extremely deforming and associated with complex squints as well as problems of dental occlusion) will then know at an early stage when various surgical procedures are likely to be undertaken. Careful follow up and accurate record keeping are facilitated, eliminating unnecessary hospital visits but ensuring that opportunities for treatment are not missed. No aspect of treatment can be fully evaluated until growth is complete. The technological revolution in diagnostic radiology—from computed tomography to three dimensional imaging and nuclear magnetic resonance scanning—is providing increasingly accurate data on which to base surgery.[6]

One aspect of facial plastic surgery creating much interest is its place in children with Down's syndrome. Surgery can reduce the size of the tongue and lower lip, augment the chin and the profile of the nasal bridge, and correct the abnormally slanting eyelids and epicanthic folds. A report that such surgery improved the social acceptance and so the development and life opportunities for a girl with Down's syndrome[7] has stimulated others to investigate its efficacy.[8-10] The treatment remains controversial. Without doubt the facial stigmas of the syndrome may be modified, but claims that speech is improved have yet to be substantiated, and some critics argue that "normalisation" of the appearance may produce new problems. Behaviour patterns which are tolerated in a child recognised as having Down's syndrome may not be if he or she looks normal.

Facial deformity may also be due to trauma or a tumour and its surgical extirpation. Craniofacial techniques are often relevant in such cases. Progress in microsurgery has spawned a plethora of new options. Soft tissue, bone, and composite

defects may be reconstructed in a single procedure with well vascularised tissue and minimal upset to the donor site. This is particularly useful for mandibular and intraoral defects but has many other applications in the head and neck. The concept of functional free muscle transfer—in which a muscle is moved to a new site, revascularised, and re-innervated by microsurgical techniques—allows dynamic reconstruction for facial palsy.[11 12] Tissue expansion—a remarkably simple idea in which a silicone bag slowly filled with saline through a subcutaneous valve is used to stretch skin—allows some defects to be filled by local tissues rather than by a distant flap.[13] It may be combined with traditional techniques. For example, if the skin of the forehead is to be used to reconstruct a large nasal defect expansion of the donor site beforehand will facilitate its direct closure, so eliminating the need for an ugly skin graft in an obvious site.

The past 15 years have seen radical developments in reconstructive methods. The full range of available skills can be offered only by specialist units, but patients, especially children, gain most if they are seen at an early stage since the timing of surgery may have a critical effect on the end result.

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Dilated cardiomyopathy, myocarditis, and the bioptome

Cardiomyopathies are defined as "heart muscle diseases of unknown cause."[1] Our lack of knowledge has acted as a catalyst to extensive research in Europe, Japan, and the United States. In dilated cardiomyopathy cases are recognised as forming a heterogeneous group probably with several aetiologies. For example, recent research in the United States has been looking at the density of β adrenergic receptors[2] and at microvascular changes[3] which result in damage of an ischaemic type with in each case a final outcome of dilated cardiomyopathy.

The infectious-immune pathogenesis has also been studied for several years. Causal relations are difficult to establish since the clinical course of dilated cardiomyopathy is far from clear. A few reports of incidence have been published giving estimates of some eight to 10 cases per 100 000 population—but these studies were based on patients with symptoms and may represent the tip of an iceberg.[4-6] Nevertheless, there is evidence to suggest that in some patients at least the likely mechanism is that a viral infection induces an immune reaction which eventually causes irreversible heart failure.

Pivotal to the clinicopathological approach has been the recovery of fresh endomyocardial tissue by bioptome. This instrument is essentially a catheter provided with a cutting device at one end, so that after standard cardiac catheterisation samples of right or left ventricle may be obtained with ease and safety.[7 8] Morphological examination of such tissue from patients clinically suspected of having dilated cardiomyopathy has shown the presence of myocarditis. The morphological diagnosis of myocarditis is difficult (as indeed is the clinical diagnosis), for the mere presence of some inflammatory cells in the myocardium is not enough: concomitant changes in the interstitium and adjacent myocardial fibres must be present before the diagnosis can be made.[9] Published reports of the incidence of myocarditis have varied between 2% and 63%.[10] This wide variation may be due to selection of patients or the underdiagnosis or overdiagnosis of myocarditis, which is often focal. Nevertheless, if five or more biopsy specimens from one or both ventricles are obtained these small tissue samples have been shown to reflect changes in the entire ventricular muscle.[11]

In 1980 an international meeting defined myocarditis as "the presence of inflammatory cells in the myocardium with evidence of fraying of adjacent myocardial fibres but without concomitant sequential fibre necrosis (as it is seen in ischaemic heart disease)."[12] Sequential biopsies in patients under treatment for active myocarditis have led to a classification into three main groups.[13] More recently a panel of (predominantly American) pathologists met in Dallas and the definition and classification were reworded, amended, and extended.[14] Depending on the relation of the inflammatory cell infiltrate, which may be sparse, to the myocardial fibres, the presence of necrosis, and the presence or absence of interstitial fibrosis the three main forms are: active, resolving (healing), and resolved (healed). This categorisation has important therapeutic implications. Patients with active myocarditis confirmed by biopsy have been treated with prednisolone and immunosuppressive agents with resolution of the inflammation and concomitant clinical improvement.[15 16] In the healing stage, too, treatment is beneficial, particularly if there are obvious foci of activity (judged by necrosis) and if the clinical outlook seems poor. If the patient is not severely disabled no more than close follow up may be needed. Once the "healed" phase is reached, however, irrespective of the clinical condition treatment by these agents is useless and if the condition is far advanced cardiac transplantation may offer the only hope.

If the first biopsy specimen shows interstitial fibrosis and foci of fibrous replacement the pathologist may surmise only that the patient had myocarditis in the past. A finding of a hypertrophied, dilated myocardium with varying degrees of fibrosis, though morphologically non-specific, may be diagnosed as dilated cardiomyopathy—provided that other causes of hypertrophy and dilatation have been ruled out.

Virological studies have shown raised antibody titres to Coxsackie B virus in patients with dilated cardiomyopathy and myocarditis, but raised titres may also be found in patients without any evidence of active myocarditis or myocarditis in the past.[17-19] Myocarditis may also be found in biopsy specimens from patients in whom the results of