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The physicochemical properties of hair have been studied from a new recessive syndrome associated with brittle hair, intellectual impairment, decreased fertility, and short stature. Electrophoresis of the SCM-structural proteins showed that the α polypeptides appeared normal, but the matrix component was markedly reduced. This was confirmed by finding a normal α x-ray diffraction pattern but a reduced ½ cystine content of hair and an abnormal stress-strain curve. Electron-microscopic studies revealed extreme disorganization of the filaments which most likely resulted from the absence of normal cross-linking. Nails, which contain structural proteins similar to hair, also showed the abnormality. Since the matrix component seen by electrophoresis consists of more than one component, the defect cannot be explained as a single structural gene abnormality.


In patients with primary gout, vascular disease tends to occur at an earlier age than in patients who do not have primary gout. This was first noted in 1899, but only in the last 25 years has evidence accumulated that hyperuricemia is a risk factor in vascular occlusive disease. Assessment of platelet ultrastructure in 124 patients with primary gout revealed abnormally increased numbers of "sticky platelets" in 55% of them. The authors' ex vivo system of platelet ultrastructure evaluation determined a direct relationship between increasing levels of uric acid and the degree of platelet stickiness. The results of the study of some of the anti-inflammatory, uricosuric, and enzyme inhibitor agents used for gout and their effect on platelet stickiness are reviewed. A drug regimen is outlined that should help to minimize the risk of platelet stickiness and hyperuricemia in primary and secondary gout.


Synthetic fluorinated derivatives of cortisone, when topically applied, are absorbed readily and may suppress adrenal function. In the case presented here, large quantities of topically applied triamcinolone acetonide were used in the treatment of atopic dermatitis. This caused a noticea-
ble decrease in the plasma cortisol level. That this was the result of adrenal suppression rather than insufficiency was indicated by a satisfactory response of the plasma cortisol level to a cosyntropin injection test. In patients with a history of recent and extensive use of topically applied corticosteroids, appropriate diagnostic and therapeutic measures are indicated to avoid postoperative adrenal insufficiency.


Technique of an alternative method for providing fixation in fractures of the jaws is presented.


Serum and dialysate levels of amikacin were determined at appropriate intervals after a 300-mg intravenous dose as a continuous infusion in six patients with end-stage renal failure undergoing hemodialysis and in three patients on peritoneal dialysis. The mean serum half-life of amikacin was 3.75 h during (or after) hemodialysis and 29 h during (or after) peritoneal dialysis. Although not on hemodialysis in the same six patients, the serum half-life was 28 h. The results indicate that the maintenance dose of amikacin should be markedly decreased in patients with severe renal failure even if they are treated with peritoneal dialysis, and that serial serum antibiotic concentrations are essential to prevent cumulative toxicity of the drug.


Lutembacher’s syndrome (mitral stenosis coexisting with an atrial septal defect) is a rare condition. Three patients with the syndrome are described, in two of whom the diagnosis was established before surgery, which was successful. The third patient died shortly after cardiac catheterization. The cardiac surgeon should look for these lesions when operating on the mitral valve or when closing an atrial septal defect.


Involutional osteopenia is the most common metabolic bone disorder affecting the elderly population. Improved diagnostic techniques now allow earlier detection of decreased bone mass than was possible with routine skeletal roentgenograms. Methods developed for quantitating bone remodeling have given insight to the mechanisms involved in skeletal loss. Current information strongly suggests that the disorder is multifactorial in origin. Theories of pathogenesis include 1) inadequate skeletal acquisition during youth, 2) dietary deficiencies of calcium and vitamin D, 3) hormonal imbalance, and 4) local factors influencing skeletal remodeling, particularly at the endosteal envelope. Numerous agents affecting bone remodeling have been used to treat involutional osteopenia, but none have been proven to restore skeletal mass. New programs involving combinations of agents give promise for increasing bone mineral content and, it is hoped, reducing fracture rate.
Abstracts


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Report of eight cases masquerading as gross aortic insufficiency, severe hypertension, myocardial infarction and mediastinal enlargement.

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