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Fig. 10.12 Morphea. Pale indurated plaques on the arm, showing a purplish rim.

fascia is not eosinophilic or permeated by eosinophils. The disease responds promptly to systemic steroids; the long-term prognosis is good but disability in the short term can be severe.

Morpheoa

Morpheoa is a localized form of scleroderma with pale indurated plaques on the skin but no internal sclerosis (Figs 10.12 and 10.13). Many plaques are surrounded by a violaceous halo. Its prognosis is usually good, and the fibrosis slowly clears leaving slight depression and hyperpigmentation. A rare type may lead to arrest of growth of the underlying bones causing, for example, facial hemiatrophy or shortening of a limb. Little is known about the cause, except that Lyme borreliosis may be associated with the disease in Europe but not in the Americas. Treatments work slowly, if at all, and include topical steroids, calcipotriene, non-steroidal anti-inflammatory drugs (NSAIDs), psoralen with ultraviolet A (PUVA), UVA and hydroxychloroquine in selected patients.

Lichen sclerosus

Many think that this condition is related to morphea, with which it may coexist. However, its patches are non-indurated white shiny macules, sometimes with obvious plugging in the follicular openings. Women are affected far more often than men and, although any area of skin can be involved, the classical ivory-coloured lesions often surround the vulva and anus. Intractable itching is common in these areas and the development of vulval carcinoma is a risk. In men the



Fig. 10.13 Morphea (en coup de sabre type) on the forehead. In a young child this can lead to facial hemiatrophy.

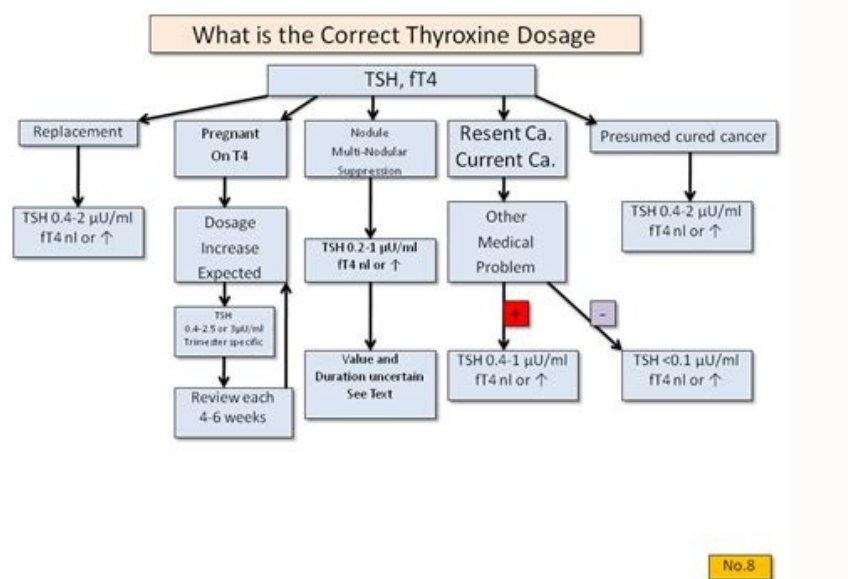
condition may cause stenosis of the urethral meatus, and adhesions between the foreskin and glans of the penis (see also Chapter 13).

Mixed connective tissue disease

This is an overlap between SLE and either scleroderma or polymyositis.

Presentation

As in LE, women are affected more often than men. Many develop swollen hands and sclerodactyly, and skin lesions like those of cutaneous LE may also be present. Alopecia is mild and the hair fall mimics telogen effluvium. Peri-ungual telangiectasia and pigmentary disturbances are common. About 25% of patients have a small vessel vasculitis with palpable purpura, leg ulcers and painful dermal nodules on the hands or elbows. Many show Raynaud's phenomenon, arthritis, serositis and myositis. Headaches, weakness, fatigue, lymph node enlargement or hoarseness occur in about one in three patients; renal and central nervous system disease are less common.



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