

Figure 1 Typical radiographs of CED patients from different families. (A) AP radiographs of both lower legs of a patient from family 14. There is cortical thickening and severe modelling defect at the diaphysis of both tibiae and fibulae. (B) Full leg radiograph (AP view) of a patient from family 2. Note the cortical sclerosis and the modelling defect with symmetrical distribution at the diaphyses of the femora, tibiae, and fibulae, with sparing of the metaphyses and epiphyses. (C ) Radiograph of the left distal femur (AP view) of a patient from family 11. Cortical thickening at the diaphysis of the femur - especially at the medial aspect - results in a modelling defect. Note sparing of the metaphysis and epiphysis. (D) Plain radiograph of the right forearm (AP view) from a patient from family 5. Cortical sclerosis and modelling defect can be seen at both radius and ulna, but are most striking at the proximal diaphysis of the ulna. (E) Standard radiograph of the forearm of a patient from family 10. Marked cortical thickening at the diaphysis of the ulna and radius can be observed, causing obliteration of the medullary cavity and hypertrophy of the long bones. Note extension of the cortical sclerosis towards the distal metaphysis of the radius. (F) Radiograph of the right arm (AP view) of a patient from family 14. Thickening of the cortex of the diaphyseal portion of the humerus, ulna, and radius is present, resulting in narrowing of the medullary canal. Note also the modelling defect of the long bones, which is most extensive at the diaphysis of the ulna. (G) Radiograph of the left hand (AP view) of a patient from family 10, showing cortical sclerosis, cortical thickening, and medullary cavity obliteration at the diaphysis of metacarpals 2 and 3. (H) Radiograph of the skull (lateral view) of a patient from family 1. Sclerosis of the calvaria, the tympanic portion of the skull base, and the ascending ramus of the mandible is visible. Note relatively small frontal and sphenoidal sinuses resulting from adjacent sclerosis of the frontal bone and upper part of the face. The maxillary sinuses are spared.