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Multiple Follicular Cysts of the Jaw 5th Phacomatosis

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The Phacomatoses (van der Hoeve, 1921) form a group of congenital, sometimes familial and hereditary, diseases in which specific disorders develop in the skin, eyes and central nervous system. They are Morbus Recklinghausen, M. Hippel-Lindau, M. Sturge-Weber and M. Pringle-Bourneville.

An eighteen year old male patient from a normal family is described. His right eye had been enucleated shortly after birth on suspicion of retinoblastoma. On renewed examination of the enucleated eye the diagnosis of dysgenesis oculoneuroblastica gliomatosa was made. At the age of three years the patient's skin began to show numerous small naevus-like lesions, distributed symmetrically over the head, neck, shoulders and body. These are still increasing in number. Diagnosis: Naevus epitheliomatodes multiplex.

Electroencephalography indicated diffuse cerebral abnormalities although intelligence appeared to be normal.

Roentgenological examination of the skeleton showed numerous abnormalities among which were cleft ribs, asymmetric and open vertebral arches and fusion of several vertebrae. The man-

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dible contained three follicular cysts not present at an earlier examination, the middle one being about three inches in diameter. A fourth cyst was seen in the left maxilla. The cysts were operated upon successfully.

The patient's and his parents' chromosomes were examined. Father and son showed the same anomaly in one chromosome.