HEBERDEN Heck

heavy chain or heavy fragment by the neoplastic cells. gamma-chain disease (y-G type and Franklin disease) is characterized by a protein antigenically related to the Fc fragment of the heavy chain of γ -G. Clinical symptoms include lymphadenopathy, splenomegaly, fever, anemia, and occasional hepatomgealy. Leukopenia and thrombocytopenia are usually present. Excessive susceptibility to bacterial infection is common. Oral manifestations include erythema and edema of the soft palate and uvula associated with Waldeyer's ring involvement. The bone marrow and lymph nodes contain atypical immature plasma cells with an admixture of atypical lymphocytes, reticulum cells, and eosinophils. The abnormal serum and urine protein show fast γ and β slow mobility; its molecular weight is about 53,000. It is immunologically related to γ-G globulin and is similar to the fast chain. The level of normal immunoglobulins is usually depressed. Alpha chain disease (a-A type) was originally described in a young Arab woman with malignant lymphoma of the intestine. It has been also observed in non-Ashkenazi Jews living in the Middle East and sometimes other ethnic and racial groups. In the original report, the abnormal protein found in serum, urine, and saliva was closely related to γ-A1 and devoid of light chains. Some normal γ-A was present; γ-G and γ-M were abnormally low. Clinical symptoms in the original case included amyloid deposits, bone pain, lymphoproliferative disorders, and carpal-tunnel syndrome, but no eosinophilia, lymphadenopathy, or recurrent infection present in other forms of heavy chain disease. MU (µ) chain disease is a rare disorder characterized by secretion into the plasma of free heavy chains. Major complaints include retroperitoneal adenopathy and hepatosplenomegaly. Lymphoplasmacytic cells may occur in the bone marrow. Most patients have large amounts of kappa-L chain in the urine. The monoclonal lymphoid cells of the neoplasm appear to have a defect in the heavy-light chain assembly.

Franklin, E. C., et al. Heavy chain disease-A new disorder of serum gamma-globulin. Report of the first case. Am. J. Med., 1964, 37:332-50.

HEBERDEN, WILLIAM, SR. (British physician, 1710-1801)

Heberden asthma. Synonyms: Elsner asthma, Rougnon de Magny disease, angina pectoris, cardiac asthma, stenocardia.

A clinical syndrome of chest pain caused by a relative oxygen deficiency in the heart muscle, occurring in the presence of arteriosclerotic coronary artery disease and, less commonly, ventricular hypertrophy, left ventricle outflow obstruction, aortic regurgitation, cardiomyopathy, and other pathological conditions of the heart. The syndrome is characterized by a distinctive radiating precordial pain, often precipitated by physical effort or emotional stress. See also Prinzmetal angina and chest wall syndrome.

Heberden, W. Some account on a disorder of the breast. Tr. Roy. Coll. Phys., London, 1772, 2:59-67.

Heberden nodes. Synonyms: Bouchard disease, Heberden syndrome, Heberden-Bouchard disease, Rosenbach disease, arthritic nodes.

Nodes produced by calcific spurs of the articular cartilage at the base of the terminal phalanges in osteoarthritis.

*Heberden, W. De nodis digitorum. In his: Commentarii de morborum historia et curatione. London, Payne, 1802. Rosenbach, O. Die Auftreibung der Endophalangen der Finger-eine bisher nocht nicht beschriebene tropische Störung. Zbl. Nervenh., 1890, 13:199-205.

Heberden syndrome. See Heberden nodes.

Heberden-Bouchard disease. See Heberden nodes. HEBRA, FERDINAND, VON (Austrian physician, 1816-

1880)

Hebra disease (1). See Gibert disease.

Hebra disease (2). See Kaposi disease (2).

Hebra prurigo. See Hebra syndrome.

Hebra syndrome. Synonyms: Hebra prurigo, von Hebra syndrome, prurigo agria, prurigo ferox, prurigo gravis, prurigo hebrae, prurigo mitis.

A chronic skin disorder that usually begins during infancy in the form of an urticarial rash followed by millet-sized or slightly larger pruritic papules which eventually become covered by a blood-colored crust. The disorder persists through life.

*Hebra, F., von. *Traite pratique de maladies de la peau*. Paris, 1854.

von Hebra syndrome. See Hebra syndrome.

HECHT, FREDERICK (American physician, born 1930) Beals-Hecht syndrome. See congenital contractural arachnodactyly syndrome.

Hecht syndrome. See trismus-pseudocamptodactyly syndrome.

Hecht-Beals-Wilson syndrome. See trismus-pseudocamptodactyly syndrome.

Hecht-Jarvinen syndrome. See popliteal pterygium syndrome.

HECHT, JACQUELINE T. (American physician)

Hecht-Scott syndrome. Synonym: limb defictency-heart malformation syndrome.

A familial syndrome, transmitted as an autososomal recessive trait, characterized by terminal transverse limb defects associated with congenital heart malformations. Acheiria, apodia, hemimelia, oligosyndactyly, and tibial bowing are the principal abnormalities of the limbs.

Hecht, J. T., & Scott, C. I., Jr. Limb deficiency syndrome in half-sibs. *Clin. Genet.*, 1981, 20:432-7.

HECHT, VICTOR (Austrian physician)

Hecht pneumonia. Synonyms: Hecht syndrome, glant-cell pneumonia, interstitial glant-cell pneumonia.

A chronic or subacute form of interstitial pneumonia occurring in infants and young children, and characterized by multinucleate giant-cell inclusion bodies

Hecht, V. Die Riesenzsellpneumonie im Kindesalter. Eine histologisch-experimentelle studie. *Beitr. Path. Anat.*, 1910, 48:263-310.

Hecht syndrome. See Hecht pneumonia.

HECK, JOHN W. (American dentist, born 1923) Heck disease. Synonym: focal epithelial hyperplasia.

Hyperplasia of the buccal, labial, and lingual mucosae, characterized by multiple, soft, sessile papules, with the lower lip seeming to be affected most frequently. First observations were made in a group of American Indian children, and later commonly found among Greenland Eskimos.

Achard, H. O., Heck, W. J., & Stanley, H. R. Focal epithelial hyperplasia found in Indian children. *Oral Surg.*, 1965, 20:201-12. Hettwer, K. J., & Rodgers, M. S. Focal