the day and evening prior to surgery. It may even be wise to wake him once during the night for that purpose.

The risk of aspiration during anesthesia precludes taking oral fluids less than four hours before surgery, but this does not affect the use of intravenous fluids. In fact, the hazards of dehydration which Holden and Maher [2] and Walters and McGowan [8] have shown do make it seem advisable to start an infusion of 5 per cent glucose in 0.45 per cent saline solution in a microdrip burette before the operation and to continue it until recovery is complete (this to be done by needle puncture of a vein, *not* by cutdown). In addition, all surgical blood loss should be scrupulously estimated by weighing the sponges, and the pharyngeal blood ooze during recovery from the anesthetic should be suctioned and measured. Any blood loss which is excessive should be replaced by a transfusion of whole blood or sedimented red cells.

Over one million sets of tonsils and adenoids are being removed in the United States each year, a rate which indicates that nearly one person in four in this country has gone through this procedure. Whether there have been appropriate medical indications for such a great number is yet another problem. The intent of this essay is to point out that 101 people are *known* to have died as a direct result of the surgery in 1964; and perhaps more have been unrecorded. The example set by Baltimore should be emulated throughout the country.

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Understanding Tay-Sachs Disease

Recent Advances

TYPICALLY becoming apparent at the age of six to eight months, Tay-Sachs disease is characterized by familial tendency, arrest of mental processes, blindness, and progressive weakness terminating in death usually at two to three years. Congenital cases have been described, as have patients in whom the disease starts in late infancy, during the juvenile period or in early adulthood. Such instances are rare, however, the bulk of reports deal with the infantile variety which as "Tay-Sachs Disease" (TSD) has been delineated from other forms of amaurotic family idiocy. Approximately 85 per cent of the patients are of Jewish extraction, but the disease has been recorded in European and American children of non-Jewish background; in babies with Asiatic and Levantine parentage, and in Negro infants.

Both in the serum and the cerebrospinal fluid the enzymes lactic dehydrogenase, fructose aldolase, and glutamic oxaloacetic transaminase are marked by elevations at the beginning of the disease and by gradual return to normal in the more protracted phases. This transient enzyme elevation has been interpreted as being parallel to the process of ganglionic destruction within the CNS. Laboratory studies, furthermore, have demonstrated increased concentrations of amino acids in the CNS, as well as elevation of the serum neuraminic acid and of the alpha-2 globulin fraction. The total serum protein and albumin tend to be decreased and the plasma gamma globulin level markedly depressed.[1]

Klenk in 1939 [4] observed that in this disease the brain contains greatly increased amounts of glycolipids for which he proposed the name "gangliosides" and that in the cerebral cortex these were present in concentrations ten to 12 times higher than normal. Subsequent workers [3, 5] have noted a marked heterogeneity of the gangliosides. By

thin layer chromatography eight components have been demonstrated. The four major gangliosides of normal human brain consist of one mono-, two di-, and one trisialoganglioside, each containing two molecules of galactose. One of the four minor gangliosides is a monosialoganglioside containing one mole of galactose and may correspond to the major component of "Tay-Sachs gangliosides." The accumulated biochemical data suggest that the Tay-Sachs ganglioside represents the unusual elaboration of a normal monosialoganglioside component which in normal cerebral gray matter constitutes less than 6 per cent of the total gangliosides.

After the second year of life most patients have marked megalencephaly, with brain weights up to 2,200 Gm. By that time the cerebral white matter displays cystic degeneration. In contrast to the cerebrum, the cerebellar hemispheres are markedly atrophic. Histochemical studies suggest that the material deposited within neurons and glial elements represents a protein-bound glycolipid which seems identical with the gangliosides.[2] Electron microscopic studies have demonstrated membranous bodies within the cytoplasm of neurons and glial cells of patients with TSD which are thought to be lysosomal in character.[10] Biochemically, these have been found to contain the gangliosides.[7]

In Tay-Sachs patients, optical microscopy shows these deposits only in the central nervous system and the neurons of the autonomic nervous system, but recent electron microscopic studies have shown lipid bodies in the liver also.[8]

TSD is transmitted as an autosomal recessive disorder and rarely occurs in successive generations. Recent observations have uncovered a deficiency of fructose-1-phosphate aldolase in sera of Tay-Sachs patients, and in 95 per cent of their parents as well.[9] These observations suggest that determination of this enzyme may help identify the heterozygous (carrier) state of TSD.—*Bruno W. Volk, M.D., Director, Isaac Albert Research Institute of the Jewish Chronic Disease Hospital; Clinical Professor of Pathology, State University of New York, Downstate Medical Center, Brooklyn, N. Y.*

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REFRESHER QUIZ

(See pages 650–651)

1. The most common inherited disease of Tropical Africa is: a. Mongolism; b. Pituitary dwarfism; c. Sickle cell disease; d. Spherocytosis.
2. Expansion of plasma volume has been reported during painful crises in sickle cell disease. *T. F.*
3. Which of the following actions are desirable in an ideal remedy for sickle cell crisis? a. break up aggregations of sickle cells; b. prevent further clumping; c. anti-sludging agent; d. each of the above.
4. Results of forcing fluid therapy in sickle cell crisis are generally encouraging. *T. F.*
5. Dramatic relief of pain in sickle cell crisis may follow intravenous vasodilators. *T. F.*