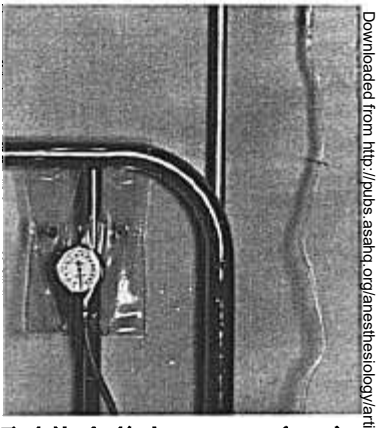
GADGET

Holder for Blood Pressure Manometer

Dr. E. W. Larking, Montreal, Canada, reports that an inexpensive plastic holder for the blood pressure gauge has been in use for several years at The Montreal General Hospital. Prior to the use of this holder, the gauges were either dropped or knocked on the floor as the patient moved in bed postoperatively. The clear plastic holder is 15 inches long, 5½ inches wide, with a 5-inch deep pocket for the blood pressure gauge. The illustration shows the holder fastened to the head of the bed by two snap fasteners with the opening for the gauge on the inside. This prevents the gauge from popping out yet it is easily visible through the clear plastic envelope. Since this holder has been in use there has been no need to replace equipment damaged by accident.



The holder for blood pressure gauge fastened to head of patient's bed.

CASE REPORTS

Refsum's Disease

Drs. Mark B. Ravin and Herman Schwartz from the Anesthesiology Service, Presbyterian Hospital, New York, report the characteristics of an unusual syndrome, heredopathia atactica polyneuritiformis, and the anesthetic management of a patient with this disease for a bilateral levator palpebrae resection for ptosis of the eyelids.

Refsum's Syndrome. In 1946 Refsum described a rare clinical neuropathy occurring in five adult members of two unrelated Norwegian families.[1] The features of this syndrome consist of progressive nerve deafness, atypical retinitis pigmentosa, ichthyosis, visual field constriction and chronic polyneuritis. Other neurologic characteristics may include diminished or absent deep reflexes, ataxia and other cerebeller manifestations, urinary sphincter impairment, loss of position sense and sensory changes involving pain and temperature. Epiphyseal dysplasia in the elbow, shoulder knee joints and marked wasting of the extremities have been noted. The cerebrospinal fluid contains a high protein content, over 200 mg. per cent, with a normal cell count. The electrocardiogram usually reveals a delayed ventricular conduction time. Myocarditis may be present.

The exact etiology of this syndrome is unknown. It appears to be a recessive hereditary characteristic since all parents or grandparents were born of consanguinious marriages.

Post mortem studies of patients with Refsum's syndrome disclosed pathologic changes resembling those of interstitial hypertrophic polyneuropathy with characteristic neural muscular atrophy, deposition of fat in affected peripheral nerves, retrograde atrophy of the ventral horns and secondary atrophy of the fasciculi gracili. In addition, a few cases demonstrated reactive changes in sympathetic ganglia, atrophy of the inferior olivary nucleus and degeneration of brain stem tracts with damage primarily to myelin. Only minor changes were noted in the extrinsic muscles of the eye and there was no clue to the etiology of the cardiac conduction defect.

The course of this disease is progressive, terminating in death after several years as a result of respiratory paralysis. No effectual treatment is known. Twenty-two cases have been reported, 14 of them children.[1-2]

Case Report. This was the first admission at the Columbia-Presbyterian Medical Center of a 13 year old Jewish Hungarian boy with a chief complaint of progressive hearing loss and ptosis of the eyelids. About two and one-half years prior to admission, there was an insidious onset of right and then left eyelid ptosis and gradual involvement of the extraocular muscles resulting in almost total ophthalmoplegia. Tinnitus had been present for one and one-half years, increasing in frequency with an associated progressive hearing loss. More recently, the patient complained of fatigue, muscle weakness and upper and lower limb incoordination. However, he denied diploplia, paresthesias, urinary sphincter difficulties, night blindness or ichthyosis.

Past History.—Birth and early growth and development were apparently normal. In November 1956, the patient was hospitalized in Vienna with viral hepatitis. In December 1956 he was re-admitted for two months with meningitis.

Family History.—Mother, father and two siblings are alive and well. The father is the off-spring of a consanguinious marriage.

Physical Examination.—The patient was well developed, but thin, with unanimated facial expression and marked bilateral ptosis of the eyelids. Blood pressure, 90/60; pulse, 80 and regular; respirations, 12 per minute; temperature, 98.8° F.; weight, 64 pounds. *Abnormal physical findings included.*—Occular: There was a marked limitation of motion of both eyes in all directions and a severe ptois of both lids. Fundoscopy revealed normal nerve heads, but the retinae appeared difusely "peppered," especially at the periphery (atypical retinitis pigmentosa). Auditory: Eighth nerve examination revealed nerve deafness more marked on the right. Nervous System: Cerebellar function examination revealed marked upper and moderate lower extremity ataxia. The motor system showed a 30 to 40 per cent decrease of strength of both upper and lower extremities. Deep tendon reflexes were decreased. Sensory examination was normal. Cardiovascular System: The heart was of normal size and heart sounds were of good quality. No murmurs were noted. Pulses of the upper and lower extremities were present and equal.

Laboratory Data: The hematocrit value was 41 per cent; blood count, urinalysis and chest roentgenograms were normal. The preoperative electrocardiogram revealed an abnormal record compatible with myocardial disease; left deviation of the electrical axis with an intraventricular conduction defect (right bundle branch block). There was regular sinus rhythm with a rate of 78. The P-R interval was 0.15 seconds. The QRS interval was 0.11 seconds and the Q-T interval was 0.36 seconds. There was a decreased response on optikokinetic tests, calorics and audiometrics. There were no abnormalities noted on electromyographic testing. The electroencephalogram showed moderately diffuse abnormalities containing paroxysmal features. On several occasions the cerebrospinal fluid had a total protein between 200 and 300 mg. per cent with gamma globulin of 8 mg. per cent. The cerebrospinal fluid pressure, sugar and cell count were normal.

Anesthetic Management. We were unable to find published reports of anesthetic management of a patient with Refsum's syndrome. Therefore, the description of the anesthetic experience with this patient may be of some interest to anesthesiologists.

The lack of clinical experience with this syndrome dictated simple straight forward anesthetic management, cautiously moving step by step. All the medical complications mentioned in the description of the disease were taken into consideration.

Small doses of thiopental were used for induction and the vital signs checked after each dose. For maintenance of anesthesia inhalation of nitrous oxide and oxygen was considered safe. Halothane was added in gradually increasing concentrations until adequate anesthesia was produced. If halothane had caused arterial hypotension or cardiac dysfunction, it would have been withdrawn rapidly. However, no change was necessary.

Summary. A patient with Refsum's syndrome (heredopathia atactica polyneuritiformis) anesthetized with general anesthesia

for bilateral resection of the levator palpebrae was discussed. The signs, symptoms, laboratory and pathologic findings of this disease were presented. Of particular concern to the anesthesiologist are progressive respiratory paralysis, a ventricular conduction defect and occasional myocarditis. Management of anesthesia for this patient with halothane-nitrous oxide-oxygen by inhalation was uneventful.

References

1. Refsum, S.: Heredopathia atactica polyneuritiformis, Acta Psychiat. Neurol., Suppl. 38: 1, 1948.
2. Refsum, S.: Hereditary atactica polyneuritiformis, J. Pediat. 35: 335, 1949; Salmoinsen, L., and Skatvedt, M.: Four cases of heredopathia atactica polyneuritiformis in children, Acta Paediat., Suppl. 77: 44, 1949; Reese, H., and Bareta, J.: Heredopathia atactica polyneuritiformis, J. Neuropath. Exp. Neurol. 9: 385, 1950; Cammermeyer, J.: Neuropathological changes in hereditary neuropathis: manifestations of syndrome heredopathia atactica polyneuritiformis in presence of interstitial hypertrophic polyneuropathy, Neurology 15: 340, 1956; Feming, R.: Refsum's syndrome: Unusual hereditary neuropathy, Neurology 7: 476, 1957; Jager, B.: Occurrence of retinal pigmentation, ophthalmoplegia, ataxia, deafness and heart block, Amer. J. Med. 29: 888, 1960.

Reaction to Thiopental

Dr. Harold A Strunk, Reading Hospital, Reading, Pennsylvania, reports a patient who reacted severely to intravenous thiopental during induction of anesthesia. It is believed that he had developed a senstitivity to thiopental, because in ten previous anesthetic procedures with thiopental there were no reactions of any kind.

The patient was a 39 year old white man admitted to the hospital on May 5, 1961, for a resection of the upper pole of the right kidney because of right renal calculus with abscess formation. Since 1944 he had had eight urological operations done under thiopental anesthesia, the last one on April 12, 1961.

The patient weighed 140 pounds, and his blood pressure was 110/50. The only other abnormal finding on examination was a Grade I systolic murmur at the apex of the heart. His blood count was within normal limits. The urine examination revealed a trace of albumin and 50 to 60 white blood cells with clumping.

The patient was given meperidine, 75 mg., and atropine, 0.4 mg. intramuscularly, one hour preoperatively. The preoperative blood pressure was 110/64; pulse, 84. At 9:59 A.M. thiopental (2.5 per cent solution) was started intravenously and 425 mg. were given. At 10:10, the patient's skin became red, no pulse was palpated and no blood pressure could be obtained. He began to vomit frequently. Oxygen was administered and a phenylephrine drip (10 mg. in 500 cc. dextrose in water) was given intravenously. At 10:40 A.M., the patient awakened and responded to questioning and did not complain of any discomfort. Hydrocortisone, 100 mg., was given intravenously at 10:45 A.M. and 12:55 P.M. His radial pulse was first palpated at 11:30 A.M., at 140 per minute, and the first blood pressure reading of 90/80 was obtained at 11:50. The pulse and blood pressure gradually improved, and at 1:50 P.M., the pulse rate was 120 and the blood pressure, 100/74. At 12:15 P.M., while still in the operating room, laboratory studies revealed a hemoglobin of 18 Gm., hematocrit of 58 per cent, white blood cells, 10,400.

When he was moved to the recovery room at 1:50 P.M., his heart was regular and his lungs were clear. He did not pass urine until the following day. A complete blood count and urine examination the next day were similar to the reports on admission. His recovery was uneventful.

On May 14, 1961, a prick test with 0.25 per cent thiopental showed a 2.5 cm. erythematous reaction. The control was negative. Skin tests with meperidine, 25 mg., and atropine, 0.4 mg., showed no local or systemic reaction. The allergist who made these tests believes that this patient had an anaphylactic-type reaction to thiopental.

The patient was again anesthetized without the use of thiopental or any other barbiturates in August 1961 for resection of the upper pole of the right kidney. No adverse reaction was observed.