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Cardiac Abnormalities during Anesthesia in a Child with Prader-Willi Syndrome

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Concurrent hypotonia, hypomentia, hypogonadism, and obesity is a description of a particular congenital disturbance, first described in 1956, and referred to as the "Prader-Willi syndrome."[1] By 1974, fewer than 200 cases had been reported. These cases demonstrate that despite a number of common characteristics, significant variability exists, particularly in growth deficiency and intellectual capability, suggesting that our atypical finding may be another expression of the Prader-Willi syndrome.

SYNDROME CHARACTERISTICS

Prader-Willi syndrome characteristics usually appear in infancy or early childhood and fall into a number of categories. Gestational abnormalities include reduced fetal activity (71 per cent), breech presentation (38 per cent), and non-term delivery (44 per cent). Decreased birth weights occur in 24 per cent of deliveries, while neonatal and infant hypotonia, prolonged feeding problems, and delayed developmental milestones occur universally.[2,3]

There is some mental retardation in virtually every patient. Afebrile seizures may appear early, but rarely are observed beyond 4 years of age. Most children who have the syndrome manifest excessive weight gain by the time they are 2 years old, although it may be delayed until 5 years of age. Short stature is visible in 95 per cent of patients,[2,3] sometimes appearing postnatally but most often not appearing until late childhood.

The facies in Prader-Willi syndrome are distinctive, typified by strabismus (48 per cent), an upward slant of the palpebral fissures, and an almond-shaped appearance of the eyes (19 per cent).[2,3] Radiographic studies of the patients demonstrate numerous craniofacial abnormalities. The anterior cranium is narrow in the bifrontal diameter in younger patients (40 per cent) and is associated with an abnormal closure pattern of the anterior fontanelle.[2-4] Hypoplasia of dental enamel and a delayed eruption pattern of the teeth constitute an occasional finding.[2,5] Following infancy the hands and feet remain small.[2]

Cryptorchidism is a common finding (89 per cent) and hypogenitalism is universal among affected males.[2,3,6,7] Evidence of hypogonadism is frequent in both male and female patients. The basis for the hypogonadism appears to be hypogonadotropism. Thirty per cent of the patients have abnormal diabetic glucose tolerance tests, but these patients are either asymptomatic or only midly symptomatic.[6,8] In addition, subnormal adrenal cortical activity has occasionally been reported.

REVIEW OF THE LITERATURE

Abnormal cardiac signs and symptoms are seen occasionally. Zellweger and Schneider were able to find one child, 5 years old, who was subject to a three-day illness characterized by sleepiness, anorexia, and dyspnea; autopsy disclosed fatty infiltration of the heart muscle and liver and bilateral pulmonary edema and pneumonitis.[7] Zellweger reports that of the 20 patients presently under his care, none has cardiovascular disease.‡ Data collected from other sources indicate cryptorchidism associated with dwarfism and diabetes in children. The diabetes has been considered a mild form, readily controlled by oral administration of hypoglycemic agents.

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Lawrence reported cases in which testicular biopsy or other operations necessitated anesthesia, but could not identify any specific anesthetic complication.§ Gellis and Feingold reported no cardiovascular or respiratory abnormality associated with Prader-Willi syndrome.[9] Hall knows of no instance of cardiac arrhythmia during anesthesia of patients with the Prader-Willi syndrome, but suggests that obese patients in general might have a higher than normal incidence.¶

Dunn, in his report of nine cases, found some isolated incidences of moderately high blood pressure and cyanotic episodes.[3]

Report of a Case

A 10-year-old boy diagnosed as having Prader-Willi syndrome was referred to the Mount Sinai Hospital after an episode of ventricular arrhythmia during anesthesia and the initiation of operation at another hospital. The operation had been cancelled and the skin incision repaired.

The child had been the product of a full-term pregnancy and normal spontaneous vaginal delivery. Birth weight was 2.64 kg. He had appeared abnormal at birth, with marked hypotonia, a large head, and a small body. He had remained hospitalized for three months, requiring extended feeding times and realizing very little weight gain. Developmental milestones had been markedly delayed; he did not walk until 4 years of age. He still has difficulty negotiating stairs, and speaks in short sentences.

The patient had weighed 2.742 kg at 12 months of age and had been hospitalized at 13 months for a pneumoencephalogram and ventriculogram to rule out obstruction. At that time he was shown to have a bone age of one month. He weighed 9.08 kg at 4 years old, but by 5½ years, he weighed 35.66 kg.

At 6 years of age, the patient had an episode of fever and cough, with development of dyspnea and cyanosis. An initial roentgenogram of the chest suggested bilateral bronchopneumonia and an enlarged heart. An electrocardiogram disclosed no abnormality. Short-term therapy with digitalis and ampicillin was initiated. The respiratory difficulties and wheezing disappeared. The pulmonary infiltrate cleared, and the heart size returned to normal. A small weight reduction accompanied hospitalization. Results of repeat glucose tolerance tests were all borderline abnormal.

Two years prior to the present admission the child was hospitalized with bilateral bronchopneumonia. The heart size was shown to be normal. Antibiotics, expectorants, and intermittent positive-pressure breathing were successful treatment.

Two months prior to admission at Mount Sinai, the patient was admitted to another hospital with a diagnosis of bilateral undescended testes associated with Prader-Willi syndrome. He was scheduled for an elective bilateral orchiopexy. During anesthesia the patient's condition was monitored with a precordial stethoscope and a display electrocardioscope only. Induction was accomplished with halothane, nitrous oxide, and oxygen through a nonrebreathing circuit. Preanesthetic pulse was 120/min. Approximately 3 or 4 minutes after incision of the skin the pulse rate increased to 180, 200, and then 250/min. The electrocardiogram did not reveal a P wave, only QRS complexes. The patient's color remained good, but the operation was cancelled. In the recovery room, 38 minutes after anesthetic induction, vital signs were: blood pressure 116/98 torr; pulse rate 120/min; respiratory rate 24/min. The initial postoperative electrocardiogram revealed sinus tachycardia with occasional premature ventricular contractions. The premature ventricular contractions disappeared by the second postoperative day.

Additional family history and genetic and metabolic studies were not helpful. The patient was obese, weighing 42 kg, and 130 cm tall. He was mentally retarded but exceptionally cheerful and communicative. Vital signs on admission were: blood pressure 120/74 torr; pulse 100/min; respiratory rate 20/min. The oral cavity had no evidence of pathologic or developmental abnormality except chronic carious lesions. The patient had poorly developed genitalia without testes in the scrotum and a recently healed inguinal skin incision.

No abnormality was disclosed by a preanesthetic electrocardiogram or by roentgenogram of the chest. Hemoglobin was 13.9 g/100 ml. Premedication two hours prior to induction of anesthesia consisted of secobarbital, 40 mg, morphine, 6 mg, and scopolamine, 0.4 mg, im. The child was cooperative but unsedated. Anesthesia was induced with high flows of nitrous oxide, oxygen, and increasing amounts of halothane added to a semiclosed circle system. Induction was monitored with a precordial stethoscope and display electrocardioscope. After light planes of anesthesia were obtained, a 19-gauge scalp vein needle and blood pressure stethoscope and cuff were placed. Initial blood pressures were 200/110 torr, with an increasingly larger number of premature ventricular contractions. The halothane concentration was increased and the trachea was intubated with the aid of succinylcholine, 60 mg, iv. The patient was then automatically hyperventilated, and in due course meperidine, 25 mg, was administered iv. The premature ventricular contractions were as frequent as bigeminy, with a total ventricular rate of 140. Blood-gas determinations and in-circuit oxygen

§ Laurance BM, Queen Elizabeth Hospital for Children, Hackney Road, London E28PS, England: Personal communication, September 30, 1974.

¶ Hall BD, Department of Pediatrics, University of California, School of Medicine, San Francisco, California 94143: Personal communication, 1975.

analysis were not available. Approximately 30 minutes after induction, blood pressure dropped to 130/100 torr and the premature ventricular contractions disappeared. The 3 per cent halothane was continued as the operation began but eventually was decreased to 1 per cent. The operation lasted 2½ hours, with one short episode of bigeminy associated with blood pressure of 160/120 torr. No specific treatment was given, and blood pressure spontaneously reverted to normal. In the recovery room vital signs were: blood pressure 120/180 torr, pulse rate 140/min; respiratory rate 32/min. Postoperative electrocardiogram in the recovery room showed infrequent premature ventricular contractions. No specific electrocardiographic diagnosis could be made. A significant hernial sac was identified during dissection of the right spermatic cord. Microscopic examination of a biopsy specimen from the right testis revealed fibrous tissue. Both testes were approximately 5 mm in longest dimension and had grossly abnormal epididymi. The child recovered from operation and anesthesia without complication.

Discussion

The patient had apparently classic symptoms and signs of Prader-Willi syndrome; however, additional findings suggested intrinsic cardiovascular abnormalities, although it cannot be determined whether such symptoms were merely extraneous complications. It seems logical that the cardiovascular abnormalities were somehow associated with the more apparent mental and physical findings of previously reported cases. They might reflect the fact that disorders of sleep, hypogonadism, obesity, psychic distrubances, sensory and motor dysfunction, disturbances of temperature regulation, diabetes insipidus, and cardiovascular abnormalities often are caused by localized defects in the hypothalamus.[2,10–12]

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