Early Prenatal Diagnosis of Conjoined Cephalopagus Twins

Imad R. Makhoul MD DSc, Dorith Goldsher MD, Marina Okopnik MD and Moshe Bronshtein MD

Departments of 1Neonatology, 2Radiology, 3Pathology and 4Obstetrics & Gynecology, Rambam Medical Center, Haifa, Israel
Affiliated to Technion Faculty of Medicine, Haifa, Israel

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One in every 50,000–100,000 births is a conjoined twin [1]. It has long been postulated that conjoined twinning is due to fission of the developing embryo. However, based on an animal model in Triton embryos and the study of 1,200 case reports of conjoined twins, Spencer [2,3] recently concluded that conjoined twinning is due to fusion of two embryos rather than fission. The union occurs either ventrally or dorsally at sites where the surface ectoderm is absent or is programmed to be fused or disrupted (primordia of heart, oropharyngeal membranes and cloacal membranes). The prognosis for conjoined twins depends on the presence of other anomalies, the extent of union of the intracranial, intrathoracic and intraabdominal structures, and abnormal vascular connections.

Patient Description

We report a case of cephalopagus diagnosed at 15 weeks of gestation by vaginal ultrasonography. After being informed of the grave prognosis of this condition, the parents opted for termination of pregnancy. Prenatal ultrasound examination (Figures A, B), MRI findings (Figure C) and a view prior to autopsy (Figure D) of the cephalopagus showed a single large head with four cerebral hemispheres, two cerebelli, two wide-spaced eyes, two ears, one nose, and one mouth with central clefts in both lips. The twins shared a cranium, ooropharynx, sternum, liver, parts of intestine and an umbilicus (two veins and two arteries). There were separate hearts, lungs, stomachs, spleens, spinal cords and female genital urinary tracts. In addition, there were four upper and four lower extremities. Our findings were compatible with cephalopagus as described by Spencer [3].

Comment

The embryonic classification of conjoined twins consists of ventral union (87%) and dorsal union (13%). There are altogether eight types of conjoined twins that vary as to the extent and location of their union [2]. The cephalopagus is the rarest type among the ventral union subgroup of conjoined twins and accounts for 11% of all cases, i.e., one in every million births [2]. It consists of a ventral union of the upper half of the body from top of head to upper abdomen. There is one large head and a wide face that results from two conjoined faces rotated through 90 degrees from the vertical axis. Hence, surgical separation of cephalopagus twins is not an option and the prognosis is extremely poor [3]. Therefore, early prenatal diagnosis of cephalopagus by either two-dimensional or three-dimensional ultrasound [4] and magnetic resonance imaging [5] are extremely important, so that termination of pregnancy can be offered to the parents as early as possible and trauma to the birth canal via vaginal delivery avoided.

During prenatal ultrasound examination, cephalopagus may be mistaken for a singleton fetus because of superposition of the conjoined fetuses and the extreme degree of fusion. Therefore, it is imperative to scan the fetus in different planes. With advanced ultrasound imaging, even earlier prenatal diagnosis of cephalopagus might become feasible in the embryo. Theoretically, cephalopagus might appear as a "V sign" by ultrasonography. Diagnosis of cephalopagus in the 7th to 8th week of
gestation will allow immediate termination of pregnancy. This is possible also for Orthodox Jews, since pregnancy termination is permitted by Jewish Law until 49 days of pregnancy.

References

Correspondence: Dr. I.R. Makhoul, Dept. of Neonatology, Rambam Medical Center, Haifa 31096, Israel. Phone: (972-4) 854-2219 Fax: (972-4) 854-3430 email: Makhoul@rambam.health.gov.il

Strychnine Intoxication in a Child
Orna Starretz-Hacham MD1,3, Shaul Sofer MD2 and Matityahu Lifshitz MD1,3
Department of Pediatrics D, Pediatric Intensive Care Unit and 3Clinical Toxicology Unit, Soroka University Hospital and Faculty of Health Sciences, Ben-Gurion University of the Negev, Beer Sheva, Israel

Key words: strychnine intoxication, poisoning, child

The use of strychnine is restricted to veterinary preparations, such as rodenticides. The tablets are a bright pink, making them attractive to children. Strychnine is rapidly absorbed following ingestion. General muscles spasm and seizures can occur a short time after intoxication and in severe cases can be fatal. We report a case of a child poisoned accidentally through ingestion of this material, who successfully responded to a muscle relaxant combined with mechanical ventilation.

Patient Description
A 6 year old otherwise healthy Bedouin boy was admitted to the Pediatric Intensive Care Unit due to severe muscle cramps. An hour before his admission he had ingested a pink tablet of unknown substance, and after approximately 30 minutes post-ingestion he began to vomit and shiver and exhibited general muscle spasms. He had found the tablets on the street near his house in a Bedouin village. These tablets are used by exterminators to rid areas of unwanted rodents and stray dogs and cats. An hour after ingestion, his parents brought the rest of the pink tablets to the PICU and the substance was identified by the exterminators as strychnine, which they had scattered a few hours before the child had found them.

At the local primary care clinic the child had been treated with repeated doses of diazepam 0.5 mg/kg/dose intravenously without any response. On admission to the PICU he was conscious but disoriented. His vital signs were: blood pressure 101/49 mmHg, heart rate 125 beats/min, temperature 37.4°C, and respiratory rate 15/min. Physical examination revealed severe, general muscle tetania and dilated pupils.

Upon admission, tests of arterial blood gases (with oxygen) showed pH 7.07, PaCO2 33 mmHg and PaO2 272 mmHg, with base excess of -20 and HCO3- 9.6 mEq/L. Blood examinations revealed hemoglobin 10.5 g/dl, white blood cell count 12,400 cells/mm3 with normal differential count, and platelet count 341,000/mm3. Urine or blood strychnine level was not measured because this test is not available in our hospital and the toxic substance had been identified.

PICU = pediatric intensive care unit

Blood analyses showed glucose 228 mg/dl and calcium 9.1 mg/dl. Renal and hepatic function were within the normal range; creatine phosphokinase was 309 U/L and after 24 hours 3,193 U/L; CPK-MB was 1.8% and myoglobin 750 mg/ml. The chest X-ray was normal and electrocardiogram showed only sinus tachycardia. The boy was intubated, mechanically ventilated, and treated at that time with a continuous drip of vecuronium bromide i.v. in a dose of 0.1 mg/kg/hour for 20 hours.

The child developed rhabdomyolysis with a peak CPK of 4,992 U/L, high serum myoglobin of 750 mg/ml, and mild renal dysfunction (creatinine 1.3 mg/dl). On the second day of hospitalization the patient developed fever and had a white blood cell count of 13,400 cells/mm3. A chest X-ray revealed an infiltrate on the left lower lobe. The sputum culture grew Streptococcus pneumoniae and he was treated with cefuroxime i.v. The boy was discharged from the PICU after 3 days in good condition; blood gases, CPK and myoglobin had returned to normal.

CPK = creatine phosphokinase