Decorticate Posture Following ‘Cardiac Cocktail’
A Transient Complication of Sedation for Catheterization

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Summary. Two children undergoing cardiac catheterization became irritable 1–3 hours after IM premedication that included phenothiazines. IV diazepam was administered and their irritability subsided. Within another 30 minutes they developed a decorticate posture from which they recovered shortly after IV diphenhydramine hydrochloride. The decorticate posture is believed to be an idiosyncratic reaction to phenothiazines. IV diphenhydramine may be preferable to diazepam when irritability develops following premedications that include phenothiazines.

Key Words: Cardiac catheterization — Phenothiazines — Decorticate posture — Diphenhydramine

Acute dystonic and extrapyramidal reactions caused by phenothiazines and related compounds in children have been reported in the literature since the late 1950s. These bizarre neurological presentations manifest mainly as torticollis, nuchal or generalized rigidity, oculogyric crises, trismus, involuntary movements, or hyperirritability[1-4, 6-10, 12]. The responsible medication is usually prescribed as an antiemetic or antitussive, but often it is accidentally ingested. To our knowledge, no cases of dystonia have been described in association with the widely used pediatric sedative “cocktail” of meperidine, promethazine, and/or chlorpromazine. Adverse reactions to “cardiac cocktail” appear as hyperirritable or hyperexcitable states or more commonly as respiratory depression with acidosis[13]. High on the list of differential diagnoses for youngsters who suddenly show abnormal neurological signs following left-heart catheterization is the possibility of an air embolus to the brain. We report an unusual neurological presentation in two children undergoing left-heart catheterization and discuss its diagnosis and management.

Case Reports

Case 1

A 3-year-old, 14.3-kg boy with congenital rubella syndrome was admitted to University of Texas Medical Branch hospitals for cardiac catheterization and cineangiography. Cardiac catheterization as a neonate revealed a large left-to-right shunt through a patent ductus arteriosus, a left-to-right shunt at atrial level, and moderate valvular pulmonic stenosis. The patent ductus arteriosus was ligated. The child has done well, although he had moderate developmental delays, especially in speech, probably associated with significant deficits in hearing and vision. He was not receiving any medications and had no history of seizures. A grade 3/6 long systolic ejection murmur was heard in the pulmonary area, and the second heart sound was widely split. The remainder of the physical examination findings were unremarkable except for bilateral ptosis and nonreactive pupils that were the result of sector iridectomies for congenital cataracts.

One hour before catheterization, the child received 1.1 ml of cardiac cocktail intramuscularly. The cocktail consisted of 25 mg of meperidine, 6.25 mg of promethazine, and 6.25 mg of chlorpromazine per milliliter. One hour into the procedure, he was given 1.0 mg of diazepam (Valium) by slow intravenous push because of increased restlessness. The child was alert and active by the end of the procedure, but within 10 minutes (2 hours 50 minutes after administration of cardiac cocktail), he suddenly became nonresponsive to verbal commands or noxious stimuli. His eyes remained open and centrally fixed, and the lids fluttered intermittently. His face was expressionless and his mouth was closed. His breathing pattern resembled Kussmaul respiration.

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Temperature, blood pressure, and heart rate remained normal. He had extreme nuchal rigidity and maintained a decorticate posture. After 5 minutes without change, he was given a further 1.5 mg of diazepam intravenously over the next ten minutes without effect. Being reasonably certain that no air had been introduced during a left ventricular injection, the possibility of an idiosyncratic or dystonic reaction to one of the components of the cardiac cocktail was considered likely. He was then given diphenhydramine hydrochloride (Benadryl) in a dose of 2 mg/kg intravenously over 5 minutes, which resulted in an immediate increase in the level of consciousness and full recovery to an alert state with normal muscle tone. Trismus was noted in the early stages of recovery. Values for serum electrolytes, calcium, BUN, and glucose drawn before the administration of diazepam and diphenhydramine were all normal. There were no recurrences of the abnormal neurological state, and he was discharged in good condition the next morning.

Case 2

A 5-year-old, 15-kg girl was admitted for cardiac catheterization and cineangiography. She had had a heart murmur from infancy diagnosed as a ventricular septal defect. The child had been asymptomatic and had no abnormal neurological developmental history. She had had no history of seizures. A grade 4/6 holosystolic murmur was heard at the third left sternal border. The heart sounds were normal, and physical examination findings were otherwise unremarkable. Thirty minutes before cardiac catheterization, the patient received 1.5 ml of cardiac cocktail. About 30 minutes into the procedure, she became agitated and received 2.5 mg of diazepam by slow intravenous infusion. About 1 hour into the procedure, 1½ hours after administration of the cardiac cocktail, and before the left-heart catheterization, the patient exhibited decorticate posture, which involved the upper extremities more than the legs and rigidity. The eyes were open and centrally fixed and her face was expressionless. Temperature, blood pressure, and heart rate were all normal. Because of our previous experience, she was given 30 mg of diphenhydramine hydrochloride intravenously, which resulted in the immediate resolution of the symptoms. She had no recurrence and was discharged in good condition.

Discussion

The differential diagnosis for this unusual neurological presentation in a child who is in good health before cardiac catheterization is limited. Meningitis, encephalitis, or tetany is most unlikely. Hypocalcemia was not present in either child. Hypoglycemia-induced seizures or coma must be seriously considered in a child who has been deprived of oral fluids and foods for a period before catheterization. These children, however, had adequate serum glucose levels and a dextrose-containing intravenous drip throughout the procedure.

Lidocaine toxicity can cause seizures, but only a small amount was used for local anesthesia. These patients’ reactions did not resemble hypersensitivity response to angiographic contrast. Hysteria is unlikely in a 3-year-old with moderate developmental delays. Children with cyanotic heart disease and polycythemia are more prone to cerebrovascular accidents, but these patients were not cyanotic and had normal hematocrit readings. A cerebral catastrophe would more likely result in this event from an air embolus during the left-heart catheterization.

In two comprehensive reviews of the complications of cardiac catheterization in children, no reference was made to idiosyncratic or dystonic reactions to phenothiazines [11, 14]. One report on another sedative mixture for cardiac catheterization referred to a “staring” episode that responded to diphenhydramine. Although the authors do not comment on it, one of the constituents was a phenothiazine-like compound, droperidol [5].

Phenothiazines are known to lower seizure thresholds in persons with epilepsy and brain damage [7]. Interestingly, anticonvulsants such as phenobarbital and diazepam can also terminate acute dystonic reactions caused by phenothiazines [10]. Both of our children had no history of epilepsy, but one does have organic brain dysfunction.

Rainer-Pope [10] recently described a group of children treated effectively for drug-induced acute dystonic and extrapyramidal reactions with diazepam, 5 to 7.5 mg intravenously. The children he treated fell asleep immediately and awoke symptom free within an hour. In the management of our patients, we hesitated to give more than 1.5 mg of diazepam. Gupta and Lovejoy [6] described a diagnostic maneuver to distinguish between seizures and drug-induced dystonia; intravenously administered diphenhydramine hydrochloride, 2.0 mg/kg over 5 minutes, caused immediate and dramatic improvement in the drug-induced dystonia. Smaller doses may fail and make the test results inconclusive. In these patients, the acute dystonic reaction occurred at about the time of peak blood levels of the phenothiazines after an intramuscular injection of cardiac cocktail. As a result of our experience, we suggest that diphenhydramine hydrochloride, 2 mg/kg given intravenously, be tried before diazepam or anticonvulsants for unusual neurological presentations at the time of or within several hours after cardiac catheterization when cardiac cocktail or phenothiazines have been employed as sedative.

These two cases should also remind those who perform cardiac catheterization using sedative cocktails that irritability in a patient may not be due to undersedation, but rather an idiosyncratic reaction to a phenothiazine-like compound in the cocktail. Additional sedation may, in fact, be hazardous.