

disorders. *In: Neurology of the Newborn*, 2nd edn. Ed Volpe JJ. Philadelphia, WB Saunders Co. 1987, pp 421.

5. McMurray WC, Mohyuddin F, Rossiter RJ, *et al.* Citrullinemia. A new aminoacid disorder associated with mental retardation. *Lancet* 1962, 1: 138-142.
6. Kennaway NG, Harwood PJ, Ramberg DA, Koler RD, Buist NRM. Citrullinemia: enzymatic evidence for genetic heterogeneity. *Pediatr Res* 1975, 9: 554-558.
7. Shannon DC, DeLong R, Bercu B, *et al.* Studies on the pathophysiology of encephalopathy in Reye's syndrome: hyperammonemia in Reye's syndrome. *Pediatrics* 1975, 56: 999-1004.
8. Msall M, Batshaw ML, Suss R. Neurologic outcome in children with inborn errors of urea synthesis: Outcome of urea-cycle enzymopathies. *New Engl J Med* 1984, 310: 1500-1502.
9. Roerdink FH, Gouw WLM. Citrullinemia: Report of a case with studies of antenatal diagnosis. *Pediatr Res* 1973, 7: 863-869.
10. Visakorpi JK. Citrullinemia. *Lancet* 1962, 1: 1357-1360.
11. Neches WH, Park SC, Ettedgui JA: Transposition of the great arteries. *In: The Science and Practice of Pediatric Cardiology*, Vol II. Eds Garson, Bricket & McNamara, Pennsylvania, Lea & Febiger. 1990, p 1174.

Cerebro Vascular Accident in Mitral Valve Prolapse

M.L. Kulkarni
A.C. Basavaraj
C. Sureshkumar
K. Farooq

Mitral valve prolapse (MVP) is a common clinical entity with a frequency in the

general population of 5-7%(1). The frequency in pediatric population is 1%(2). MVP is the most common cardiac abnormality observed echocardiographically in stable newborn girls(3).

The vast majority of patients with MVP have no serious complications. However, there are a few major complications, that may accompany MVP, they are: chest pain, progressive mitral regurgitation, infective endocarditis, thromboembolism, serious arrhythmias and sudden death(4).

Thromboembolism leading on to cerebrovascular accidents (CVA) is occasionally reported in adults(5). But its occurrence in pediatric age groups is exceedingly rare and no such cases have been reported in the literature, that too, from a clinically silent MVP. We report such a case.

Case Report

A 10-year-old girl was admitted with history of sudden onset of weakness in right upper and lower limbs associated with inability to talk, 6 months prior to admission.

Physical examination revealed that her anthropometric measurements were normal for her age. Her pulse was 80 beats per minute and regular, BP was 100/70 mm of Hg. All her peripheral arterial pulsations were well felt. She had no neurocutaneous markers or cutaneous changes seen in collagen vascular disorders. Central nervous system examination revealed that she had expressive aphasia, right sided upper

From the Department of Pediatrics, JJM Medical College, Davangere 577 004, Karnataka
Reprint requests: Prof. M.L. Kulkarni, 2373, M.C.C. 'A' Block, Davangere 577 004, Karnataka.

Received for publication: February 11, 1992;
Accepted: November 20, 1992

motor neuron type of facial weakness with dense hemiplegia on the rightside. Her ocular fundii were normal. Other systems including cardiovascular system were essentially normal on clinical examination.

Her Hb was 9.2 g/dl, total blood count 7,000 cells/cu mm, ESR 13mm/hour, platelet count 1.5×10^5 cu mm, serum cholesterol 130 mg/dl, blood urea 19 mg/dl, random blood sugar 80 mg/dl and prothrombin index was 100%. The routine urine examination did not reveal any abnormality. Tests for LE cell phenomenon, rheumatoid factor, sickling phenomenon, PPD, BCG test, ECG, skull and chest X-ray were all normal. An echocardiogram revealed that the child had typical features of mitral valve prolapse which showed sagging of mitral valve leaflets in both four chamber and long axis view in 2-D Echocardiogram.

Discussion

The typical patient with MVP is a young woman of tall stature, long arm span and lean built with narrow anteroposterior chest diameter. About 42% of the patients are asymptomatic, in addition 5 to 7% of patients have silent mitral valve prolapse with no audible murmurs or clicks(5). The discovery of one or more midsystolic clicks with or without late systolic murmur at the apex is usually incidental or accidental. The most frequently reported symptoms have been palpitations, chest pain, dyspnea, fatigue, lassitude, anxiety, neuropsychiatric symptoms, hyperventilation and syncope.

Thromboembolism is a rare complication of MVP but a sixfold increased incidence of MVP is seen in young patients who had a stroke, over controls(5). Emboli in a case of MVP may result from fissuring and thrombus formation on valves, or from the formation of aggregates of platelets

and fibrin in the angle between the posterior mitral leaflet and the left atrial wall. Abnormal platelet coagulant activities, shortened platelet survival time and plasma platelet hyperactivity have been postulated as mechanisms relating to emboli from the prolapsing mitral valve(4).

The entity acute infantile hemiplegia seen in pediatric practice is a disorder that affects infants and children under two years of age(7). The episode of cerebrovascular accident in the present case with an underlying MVP makes one think of the association between the two entities. The studies showing many fold increased incidence of MVP in young patients who had cerebrovascular accident gives circumstantial evidence that links, cerebral embolism to mitral valve prolapse(5). Not a single case among 50 and 30 children with MVP had any complications in two separate studies(2,6). In a large long term follow-up study, only 10 among 237 patients with MVP developed cerebrovascular accident and none of them were in the pediatric age group, though the study included patients in the age range of 10-69 years. In that large group there were 20 silent MVP cases and none of them had any complications(8). The present case is the first case of silent MVP with cerebrovascular accident in pediatric age group reported in the literature.

A child who presents with cerebrovascular accident with no evidence of precipitating factors, and a clinically normal heart, should undergo detailed evaluation of cardiovascular system including echocardiogram to detect silent MVP, which may be the cause of embolic phenomenon. Most cases of mitral valve prolapse are managed conservatively. In asymptomatic persons prophylactic antibiotics are recommended

in circumstances likely to produce bacteraemia. Antithrombotic and antiplatelet therapy such as aspirin and dipyridamole should be considered in patients with cerebral embolic events. How long anticoagulants should be continued is unknown(5).

REFERENCES

1. Procacci PM, Savran SV, Schreiter SL, *et al.* Prevalence of clinical mitral valve prolapse in 1169 young women. *N Engl J Med* 1976, 294: 1086-1088.
2. Shamburger RC, Welch KJ, Sanders SP. Mitral valve prolapse associated with pectus excavatum. *J Pediatr* 1987, 3: 404-406.
3. Chandraratan PAN, Vlachovich G, Kong Y, Wilson D. Incidence of mitral valve prolapse in one hundred clinically stable new born baby girls. An echocardiographic study. *Am Heart J* 1979, 98: 312-314.
4. Cheng TO, Mitral valve prolapse. *Dis Mon* 1987, 33: 483-534.
5. Barnet HJM, Boughner DR, Taylor DW, Cooper PE, Kostuk WJ, Nichol PM. Further evidence relating mitral valve prolapse to cerebral ischemic events. *N Engl J Med* 1980, 302: 139-144.
6. Sukumaran TU, Manjooran RJ, Thomas K. A clinical profile of mitral valve prolapse syndrome. *Indian J Pediatr* 1990, 57: 771-773.
7. Dooling EC. Acute hemiplegia in infancy. *Indian J Pediatr* 1990, 57: 325-335.
8. Nishimura RA, McGoon MD, Shub C, Miller PA, Ilstrup DM, Tajik J. Echocardiographically documented mitral valve prolapse. Long-term follow-up of 237 patients. *N Engl J Med* 1985, 313: 1305-1309.

Neurological Assessment at Three Months as a Predictor for Developmental Outcome in High Risk Infants

Sudha Chaudhari
Sujata Kulkarni
Anand Pandit
U.K. Koundinya

With modern methods of neonatal care, babies have begun to survive after insults, which were previously thought to be fatal. As a result of this, there is an increase in the number of survivors and also in the type of brain lesions from which they suffer(1). Hence, it is becoming increasingly important to keep a close watch on the neurodevelopment of these graduates of the neonatal intensive care unit (NICU).

Ideally, all high risk babies need close surveillance throughout infancy. However, in developing country like ours, with limited resources, a poor transport system and rising cost of fuel, this is not possible. It is essential to identify a group of babies, early on in infancy, who do not need close surveillance as far as neurodevelopment is concerned. A study was undertaken to determine if a 3 month neurological assessment could be used to predict the neurodevelopmental outcome at one year. This

From the Division of Neonatology, Department of Pediatrics, K.E.M. Hospital, Pune 411 011.

Reprint requests: Sudha Chaudhari, Consultant, Division of Neonatology, Department of Pediatrics, K.E.M. Hospital, Pune 411 011.

Received for publication: July 10, 1992;

Accepted: December 24, 1992