Cyanotic congenital heart disease: Our constraint in a resource-limited setting

Sir,

Cyanotic congenital heart disease constitute a sizable population of our patients with congenital heart disease, and tetralogy of Fallot is the commonest cyanotic heart disease seen in pediatric cardiology clinic of most centers in developing countries for obvious reason, because it is the commonest congenital cyanotic heart lesion that sufferers may survive into late childhood and adulthood without surgical intervention; lack of centers with expertise in pediatric cardiac surgery has condemned those affected with defects like transposition of the great arteries, tricuspid atresia and the likes to early death. Therefore, clinical observation of most of the complications of uncorrected cyanotic congenital heart disease is often seen in our setting. Though we have managed several children with these problems but the index case is the oldest patient and had been lucky to have had modified Blalock-Tussig (B-T) shunt and had several complications of cyanotic congenital heart disease over the years from repeated hyper-cyanotic spells, through thrombocytopenia to pediatric stroke. Therefore, I wish to highlight our experience with an 11-year-old boy being following for tetralogy of Fallot in a Federal Medical Centre with limited facilities for cardiac care in Nigeria. He was diagnosed with tetralogy of Fallot at the age of 8-month based on the complaint of cyanosis and recurrent fast breathing which were tet spells. He has been regular with clinic visits despite that, at about 3rd year of illness he developed thrombocytopenia with iron deficiency and at this point he had massive hematochezia warranting blood and platelet transfusion. After recovery parents were able to the source fund for surgery abroad. Despite modified B-T shunt, his hematocrit had ranged between 45% and 65% and at the age of 8 years he had febrile illness with right-sided hemiparesis; he was treated for cerebral abscess with stroke. He made remarkably improved on antibiotics and physiotherapy. He is currently awaiting definitive surgery.

Prevalence of thrombocytopenia in congenital cyanotic heart disease is not known, and decreased platelet production due to right to left shunting of megakaryocytes away from the lung has also been implicated as a possible cause. Furthermore, they are prone to infective endocarditis and coupled with right to left shunting of infected thrombi which may embolize to body organs including the brain resulting in cerebral abscess and mycotic aneurysm. His hematocrit still remained high despite the B-T shunt because the B-T shunt was now inadequate for his age. Availability of definitive surgery in our setting will certainly improve his quality of life and survival.

Ibrahim Aliyu, Zainab F. Ibrahim
Department of Paediatrics, Aminu Kano Teaching Hospital, Bayero University, Department of Nursing, Aminu Kano Teaching Hospital, Kano, Nigeria

Address for correspondence: Dr. Ibrahim Aliyu, Department of Paediatrics, Aminu Kano Teaching Hospital, National Postgraduate Medical College of Nigeria, Kano, Nigeria. E-mail: ibrahimaliyu2006@yahoo.com

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