

# Congenitally corrected transposition of great arteries with ischemic symptoms in middle age

Syed M. Zaidi, MD, MRCP, Mabrooke M. Al-Sharary, MD, Tarig S. Al-Khuwaitir, ABIM, MRCP, Najeeb U. Sajid, MBBS, DIP CARD.

## ABSTRACT

Congenitally corrected transposition of the great arteries (CCTGA) is a rare congenital disease first described by Von Rokitansky in 1875. Transposition of the great arteries comprises 2.6 - 7.8% of all cases of congenital heart disease, and if uncorrected, is commonly fatal in the first year of life. Patients with corrected transposition of the great arteries without associated defects may remain undiagnosed until adult life. Symptoms occur rarely before the fourth and fifth decades, when rhythm disturbance, left atrioventricular valve regurgitation, and moderately impaired systemic ventricular function cause congestive cardiac failure. We report here a case of drug overdose with ischemic symptoms, and CCTGA without associated anomalies in a 40-year-old male.

*Saudi Med J 2007; Vol. 28 (10): 1597-1599*

*From the Departments of Cardiology (Zaidi, Al-Sharary, Sajid), and Internal Medicine (Al-Khuwaitir), Riyadh Medical Complex, Riyadh, Kingdom of Saudi Arabia.*

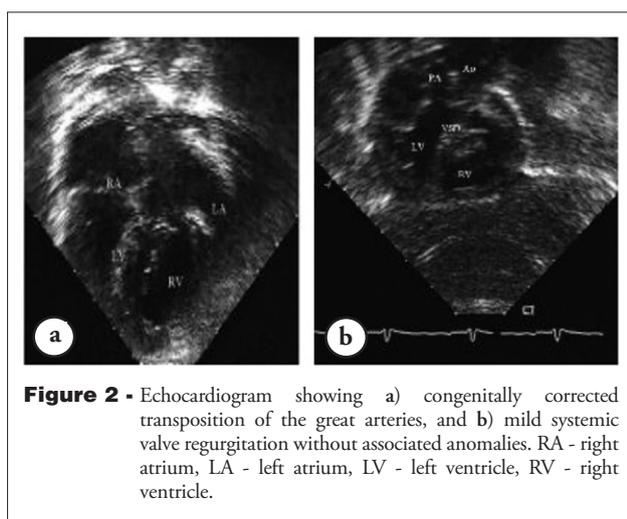
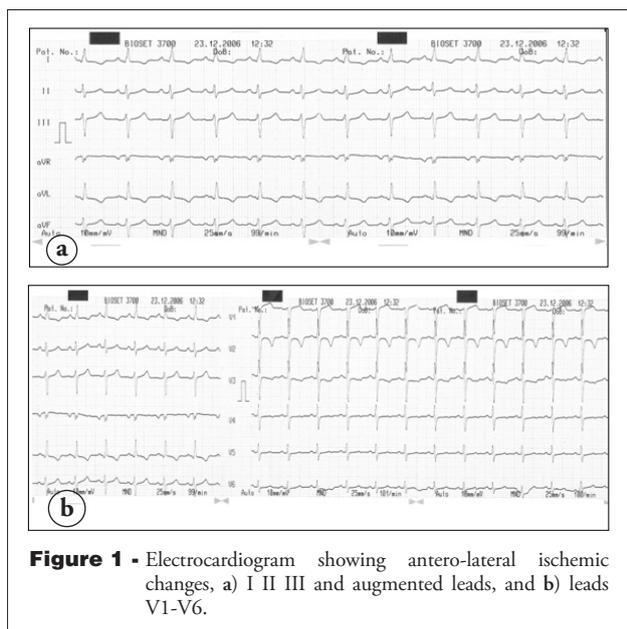
*Received 5th September 2006. Accepted 10th January 2007.*

*Address correspondence and reprint request to: Dr. Syed M. Zaidi, Department of Cardiology, Riyadh Medical Complex, Ministry of Health, PO Box 57356, Riyadh 11574, Kingdom of Saudi Arabia. Tel. +966 (1) 4511613. Fax. +966 (1) 4783446. E-mail: syedmihdi5@hotmail.com*

Congenitally corrected transposition of the great arteries (CCTGA) is a rare congenital disease first described by Von Rokitansky in 1875.<sup>1</sup> It is characterized by atrioventricular (AV) and ventriculoarterial discordance. The physiologically corrected circulation is with the morphologic right ventricle (RV) serving as the systemic pump, which is the result of this double discordance.<sup>2</sup> Patients with CCTGA (AV and ventriculoarterial discordance) are a heterogeneous group, often having ventricular septal defects, AV valve abnormalities, semilunar valve abnormalities, tetralogy-type malformations including pulmonary atresia, and

twisted or hypoplastic ventricles.<sup>3</sup> Development of the epicardial and myocardial coronary arteries involves the predictable migration of developing vessels along the planes of the AV and interventricular grooves.<sup>4,5</sup> Abnormalities of coronary origin and proximal course are seen, such as common mixed trunk (single coronary artery), and vessels crossing the RV outflow tract. Beyond its origin and proximal course, the morphology of the coronary artery was observed to “follow” that of the ventricle.<sup>6</sup> We report here a case of myocardial ischemia, due to drug overdose, in CCTGA without associated anomalies.

**Case Report.** A 40-year-old “drug trafficker” was presented with history of swallowing a large number of tablets, with the aim of concealing the evidence. He was uncooperative in volunteering any detailed history. Except for mild chest discomfort, there was no symptoms of abdominal pain, vomiting, diarrhea, shortness of breath, or loss of consciousness. No history of smoking, diabetes mellitus (DM), hypertension (HTN), dyslipidemia, or ischemic heart disease (IHD) in the family. No past history of hospitalization due to any cardiac problem. There was no family history of congenital heart disease, DM, HTN or chronic illnesses. On physical examination, he is of normal build, conscious and oriented with agitated mood, but not in distress. He was not pale, cyanosed, or jaundiced. No lymph node was palpated. His temperature was normal, pulse rate 82/min, blood pressure 150/100 mm Hg, and normal jugular venous pressure. The cardiac auscultation revealed a single, second heart sound and pan-systolic murmur at the apex. The examinations of other systems were essentially normal. He was admitted with an impression of drug overdose, and treated with gastric lavage and charcoal. His hemoglobin, blood sugar, urea, creatinine, and electrolytes were within normal range. The toxicology screen was positive for amphetamine. His creatinine kinase was elevated (328 u/l, normal range: 25-195 iu/L), and the electrocardiogram showed



antero-lateral ischemic changes (Figure 1). The thallium scan showed antero-septal perfusion defect at rest. The echocardiogram (Figure 2), showed CCTGA and mild systemic valve regurgitation without associated anomalies with mild anterior wall hypokinesia, with an overall ejection fraction of 45%. He was referred to the cardiac catheterization laboratory for coronary angiography that showed anomalous origin of coronaries, left circumflex and right coronary artery arising from left coronary sinus, and left anterior descending coronary artery from right coronary sinus, without any obstructive lesion. Hence this patient, who took over dose of amphetamine, developed acute coronary syndrome, non-ST elevation myocardial infarction. For this he was treated in the hospital, and was discovered to have CCTGA.

**Discussion.** Transposition of the great arteries comprises 2.6 - 7.8% of all cases of congenital heart diseases, and if uncorrected, is commonly fatal in the first year of life.<sup>7</sup> The clinical course is complicated by associated intracardiac defects such as ventricular septal defect, subvalvar and valvar pulmonary stenosis, left AV valve regurgitation, and AV conduction disturbances.<sup>8</sup> Patients with corrected transposition of the great arteries without associated defects may remain undiagnosed until adult life. Symptoms occur rarely before the fourth and fifth decades, when rhythm disturbance, left AV valve regurgitation, and moderately impaired systemic ventricular function cause congestive cardiac failure.<sup>9</sup> This is supported by another study that patients with CCTGA are increasingly subject to congestive heart failure (CHF) with advancing age; this complication is extremely common by the fourth and fifth decades. Tricuspid (systemic AV) valvular regurgitation is strongly associated with RV (systemic ventricle in CCTGA) dysfunction and CHF, whether it is causative or a secondary complication remains speculative.<sup>10</sup> Little attention has been given to myocardial perfusion and the possible role of myocardial ischemia, infarction, or both, in right ventricular dysfunction.<sup>11</sup> Impaired myocardial blood flow reserve of the anatomic RV in the absence of ischemic symptoms may be associated with reduced ventricular function of the systemic chamber.<sup>12</sup> As discussed previously, a patient with CCTGA usually presents with rhythm disturbance, left AV valve regurgitation, and moderately impaired systemic ventricular function causing congestive cardiac failure. Symptoms occur rarely before the fourth and fifth decades.<sup>9</sup> We conclude that in our case, the 40-year-old man, in whom we discovered CCTGA without intracardiac anomalies, presented with symptoms of angina due to vasospasm, possibly due to drug overdose of amphetamine.

## References

1. Von Rokitsansky K, editor. Die Defected der Scheidewande des Herzens: Anonymous Pathologische-anatomische Abhandlung. Wien (Vienna): Wilhelm Braumuller; 1875. p. 83-86.
2. Dyer K, Graham TP. Congenitally corrected transposition of the great arteries: current treatment options. *Curr Treat Options Cardiovasc Med* 2003, 5: 399-407.
3. Mawson JB. Congenital heart defects and coronary anatomy. *Tex Heart Inst J* 2002; 29: 279-289.
4. Vrancken Peeters MP, Gittenberger-de Groot AC, Mentink MM, Hungerford JE, Little CD, Poelmann RE. The development of the coronary vessels and their differentiation into arteries and veins in the embryonic quail heart. *Dev Dyn* 1997; 208: 338-348.
5. Vrancken Peeters MP, Gittenberger-de Groot AC, Mentink MM, Poelmann RE. Smooth muscle cells and fibroblasts of the coronary arteries derive from epithelial-mesenchymal transformation of the epicardium. *Anat Embryol (Berl)* 1999; 199: 367-378.

- 6 Shea PM, Lutz JF, Vieweg WV, Corcoran FH, Van Praagh R, Hougen TJ. Selective coronary arteriography in congenitally corrected transposition of the great arteries. *Am J Cardiol* 1979; 44: 1201-1206.
- 7 Liebman J, Cullum L, Belloc NB. Natural history of transposition of the great arteries: anatomy and birth and death characteristics. *Circulation* 1969; 40: 237-262.
- 8 Ikeda U, Furuse M, Suzuki O. Long term survival in aged patients with corrected transposition of the great arteries. *Chest* 1992; 101: 1382-1385.
- 9 Presbitero P, Somerville J, Rabajoli F, Stone S, Conte MR. Corrected transposition of the great arteries without associated defects in adult patients: clinical profile and follow-up. *Br Heart J* 1995; 74: 57-59.
- 10 Graham TP Jr, Bernard YD, Mellen BG. Long-term outcome in congenitally corrected transposition of the great arteries: a multi-institutional study. *J Am Coll Cardiol* 2000; 36: 255-261.
- 11 Hochreiter C, Snyder M, Borer J, Engle M. Right and left ventricular performance 10 years after Mustard repair of transposition of the great arteries. *Am J Cardiol* 1994; 74: 478-482.
- 12 Bengel HM, Hager BF, Kuehn A. Impaired myocardial blood flow and coronary flow reserve of the anatomical right systemic ventricle in patients with congenitally corrected transposition of the great arteries. *Heart* 2003; 89: 1231-1235.

## Case Reports

Case reports will only be considered for unusual topics that add something new to the literature. All Case Reports should include at least one figure. Written informed consent for publication must accompany any photograph in which the subject can be identified. Figures should be submitted with a 300 dpi resolution when submitting electronically or printed on high-contrast glossy paper when submitting print copies. The abstract should be unstructured, and the introductory section should always include the objective and reason why the author is presenting this particular case. References should be up to date, preferably not exceeding 15.