

CASE REPORTS

Left Atrial Rhythm in Cyanotic Congenital Heart Disease

by

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(Report of two cases, one with DORV, and one with mirror image dextrocardia associated with TF).

Abstract

Left atrial rhythm was very rarely reported in the literature, probably because of its rare occurrence. Two cases in our center that fulfilled the criteria proposed by various authors abroad were reported. One case was associated with Double Outlet Right Ventricle, Taussig-Bing type, while the other was associated with concomitant mirror image dextrocardia and Tetralogy of Fallot.

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Introduction

Left atrial rhythm is an extremely rare ectopic atrial mechanism with slow rhythm, which originates from an ectopic pacemaker in the left atrium (Chung, 1971b). The way of impulse in the atria is from the left atrium to the right atrium, instead of from right to left as found in the usual sinus rhythm which originates from the sinus node pacemaker. In normal circumstances the cardiac activation arises in the head of the sinus node, and the relation of the right atrium to the left atrium determines the direction of the P wave in lead I: the P wave is upright in normal subjects and in dextroversion of the heart, while the P wave is negative when the atria are inverted as in mirror image dextrocardia. One of the exceptions to this rule is ectopic left atrial rhythm (Mirowski et al., 1962, 1963; Somlyo and Grayzel, 1963; Nadas and Fyler, 1973; El-Sherif, 1970; Waldo et al., 1970).

The direction of the P wave in lead I has generally been considered as diagnostic for the position of the atria. When the direction of the P wave in lead I is opposite to that expected from the relation of the atria, the possibility of left atrial rhythm should be considered (Mirowski et al., 1963; Somlyo and Grayzel, 1963). Indeed, the ectopic slow atrial rhythm has received very little attention until recently because of its relatively rare occurrence. Nevertheless, Lewis had studied the subject as early

as 1910. In his experiments different configurations of the P waves were observed, depending on the location of the stimulation given. The P wave was found to be upright in lead II when the upper zone of the atria of a thoracotomized dogs was stimulated, and inverted when the lower zone was stimulated. Abramson et al. (1938) found that lead I was most useful for distinguishing between the left and right atrial rhythm. In his experiment the P wave was inverted in lead I when the left atrium was stimulated, while it was upright when the right atrium stimulated

Regarding the left atrial rhythm, Mirowski et al. (1963, 1964, 1966) mentioned that beside the abnormal direction of the P wave in lead I, there was another specific finding in all of his patients with left atrial rhythm, i.e. an abnormal configuration of P wave in right precordial leads facing the right atrium. These abnormal P waves were called "dome and dart" P waves, characterized by a notched upright wave with an initial smooth low voltage deflection, and followed by a sharp higher peaked second component. But, subsequently, it was found to be a relatively uncommon (Harris, 1968; Mirowski, 1966; Chung, 1971b). Such P waves were observed when the pacemaker was ectopic and were not seen during sinus rhythm. The dome is produced by the left atrium, which is activated first, and the dart component represents late activation of the right atrium. When there is concomitant left atrial enlargement the ampli-

tude of the first component will be increased to more than 0.2 mV, and the peak interval (the distance between the summits of the 2 components) will be more than 0.05 second; while in cases with right atrial enlargement the voltage of the dart component will be increased, reaching more than 0.45 mV.

Another important thing to diagnose left atrial rhythm is inverted P wave in the left precordial leads, at least V6, as described by Mirowski (1966) and Chung (1971). Costant (1973) mentioned that the rare "dome and dart" P wave is the only certain sign of a left atrial rhythm; while Friedman (1971) described that criteria for the diagnosis of a left atrial rhythm consist of 3 electrocardiographic characteristics: (a) inverted P waves in lead I, (b) dome and dart P wave in lead V1, and (c) inverted P wave in lead V6. Chung (1971) made a conclusion that the diagnosis of the left atrial rhythm is certain only when the ECG shows inverted P waves in leads I and V6, dome and dart P wave in lead V1, inverted P waves in leads II, III, aVF and upright P wave in aVR. However, as long as the P wave is inverted in V6 and upright in V1, in addition to being inverted in leads II, III, aVF, the diagnosis of left atrial rhythm can be made with reasonable certainty. Inverted P waves in lead I and dome and dart P wave in lead V1 are not essential for the diagnosis of left atrial rhythm. Measurement of the P-R interval or the cardiac rate is not helpful in diagnosing left atrial rhythm.

The clinical significance of left atrial rhythm is still not clearly evident, but it has been observed in apparently healthy individuals as well as in patients with diseased hearts (congenital and rheumatic heart diseases). Left atrial rhythm does not seem to be related to any cardiac drugs, including digitalis, nor it is reported to occur in acute myocardial infarction (Chung, 1971b). According to Mirowski et al. (1963), it was usually possible to convert a sinus rhythm to an ectopic rhythm and vice versa. The valsalva maneuver was frequently effective; changes in rhythm were also obtained by applying ocular pressure or by exercising the patient. The new rhythm obtained by these maneuvers was transient, usually lasting only a few seconds or minutes. In the report of Mirowski et al (1963), it was mentioned that all his patients with diseased hearts associated with left atrial rhythm tended to suffer from frequent spontaneous or induced supraventricular arrhythmias.

It has been described by various authors that the left atrial rhythm, especially with dome and dart P wave, was very rare and seldom reported in the literature. But, however, in its presence one might anticipate and should be prepared against the possible induced supraventricular arrhythmias, especially during catheterization and angiocardiology. Those are the reasons for presenting this case report.

Figure 1 Case 1 Rel. Az., 5 years



A. Left atrial rhythm (1977)

B. Atypical sinus rhythm with left-axis deviation (1977)

C. Left atrial rhythm (1977)

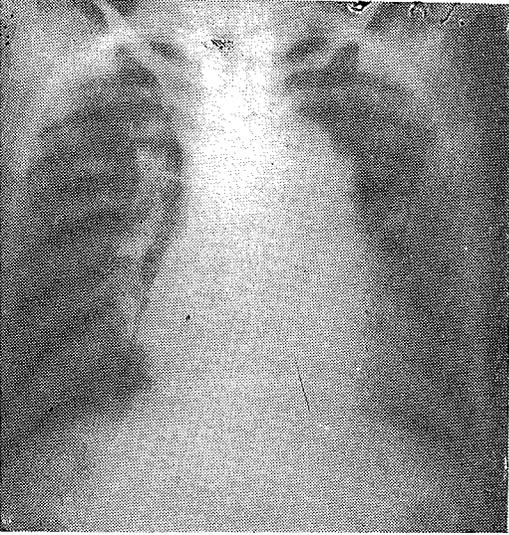


Fig. 2A



Fig. 2B

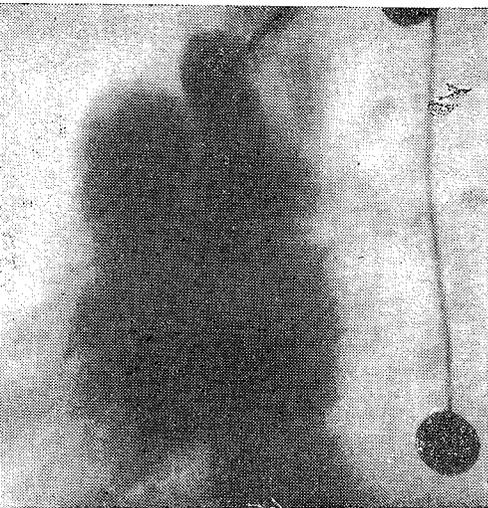


Fig. 3A

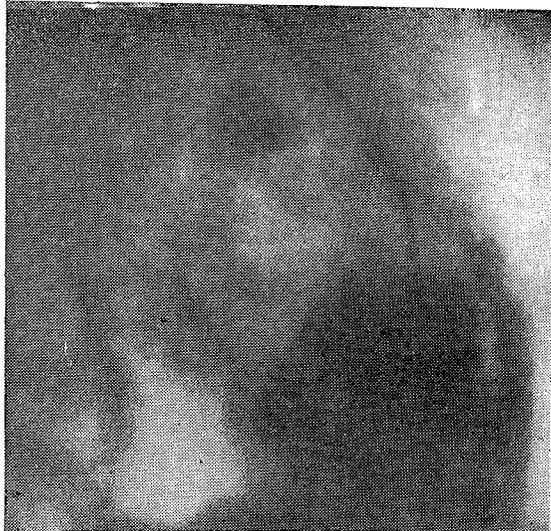


Fig. 3B

Case report

* Case 1

Ach. A., a 5-year-old Indonesian boy, was referred to the Cardiac Clinic for the diagnosis of cardiac abnormality. The family history was non-contributory. His mother never had German measles nor any rash or unexplained fever during her pregnancy. He was the third of 5 children in the family. The past history indicated that Cyanosis was noticed at early infancy and persisted, but the parents considered the abnormality as not urgent to bring the child to a competent physician. His development was said to be slow; he managed to sit by himself at 9 months and began to walk at 2 years. He had had recurrent bouts of respiratory infection. At about 3 years of age he frequently squatted down to rest, but later on outgrew the habit. Several days previously he was seen by a physician for a respiratory infection, and it was then for the first time a heart murmur was heard.

Physical examination

The temperature was 37° C, the pulse rate 120/minute, respiration rate 30/min., weight 13 kg., height 95 cm., and blood pressure 90/60 mmHg. The child was an alert, moderately cyanotic, poorly developed boy who suffered from mild dyspnea at rest. The lips and buccal mucous membranes were cyanotic. The tonsils were slightly enlarged. The teeth were in fair condition; the

chest was slightly asymmetric with bulging on the left side. The heart was slightly enlarged with increased right and left ventricular activity. The rhythm was regular. A systolic thrill was palpable and a loud systolic murmur was heard over the III-IV LSB and suprasternal notch; a diastolic murmur was heard on the apex; the second heart sound closely split with loud pulmonary component. The lungs were clear to percussion and auscultation. The liver and spleen were not palpable. The pulse in the femoral artery was of good quality. There was cyanosis and clubbing of the fingers and toes.

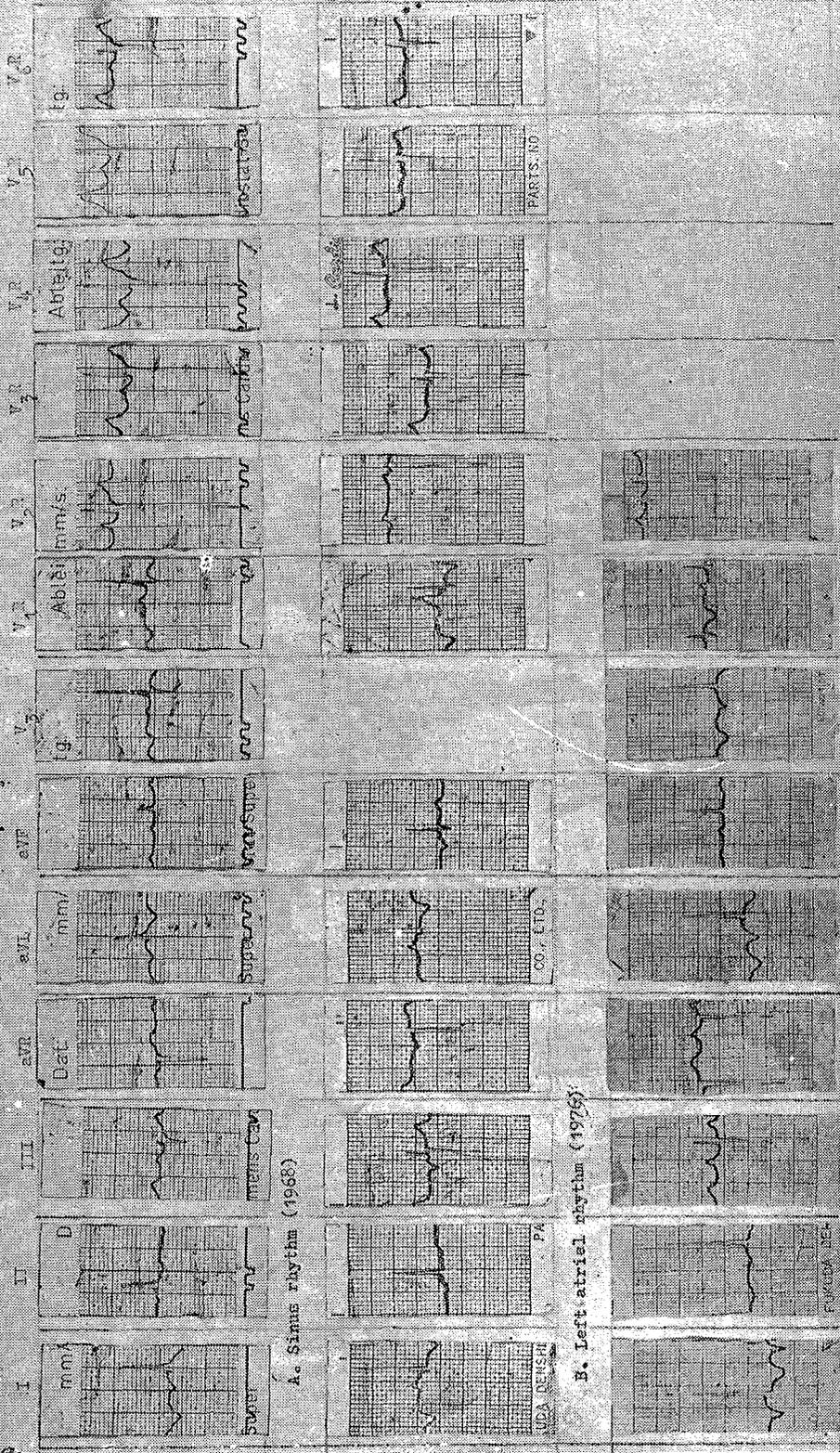
Laboratory data

Hemoglobin concentration was 16.6 gm.%; erythrocyte count 5.4 million/cum; hematocrit 53%; thrombocyte count 314,000/cumm; and leucocyte count 9,000/cumm. Urine was normal. Chest X-ray examination revealed a normally placed heart, but rather slightly enlarged; there was prominence of the pulmonary conus, and lung vascularity was increased indicating a left to right shunt. Lateral view showed enlargement of the left atrium and right ventricle (Fig. 2).

Electrocardiogram

P waves inverted in leads I, II, III, aVF, V5 and V6; upright in aVR; done and dart P wave in V1 with wide and tall first component; P axis was -140° / $+220^{\circ}$. PR interval was 0.16"; QRS axis $+130^{\circ}$, QRS rate 120, slurring of QRS in some leads deep S in V1 V2,

Figure 4; Case II, St. Pa., 0, 13 years



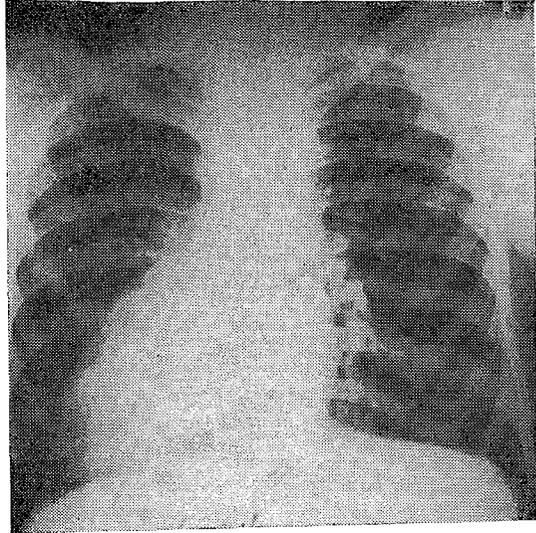


Fig. 5A



Fig. 5B

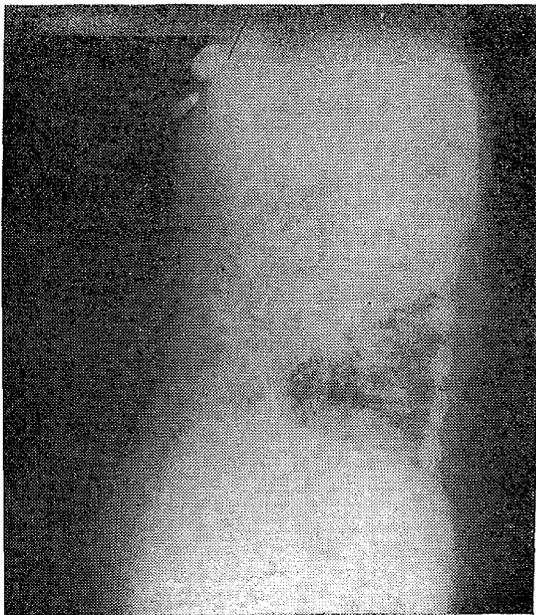


Fig. 6A



Fig. 6B

deep S in V5. ECG diagnosis of the left atrial rhythm and LAH, and suggestive CVH was made.

Hemodynamic studies

Catheterization and angiocardiography were done 3 times. The first 2 studies could not be completed due to frequent outbursts of atrial tachycardia. The last study with right and left heart catheterization and angiocardiography successfully revealed the real anatomical lesion: Double Outlet Right Ventricle (DORV), S.D.D. type, with supracristal/subpulmonary Ventricular Septal Defect, without pulmonary stenosis (Taussig Bing type). In this patient, the left atrial rhythm almost persisted. Catheterization and angiocardiography induced atrial tachycardia which then returned to the left atrial rhythm. But during observation, the ECG sometimes spontaneously showed atypical sinus rhythm with extreme left P axis deviation (with inverted P in II, III, aVR, aVF, and normal P-R interval), and then again spontaneously returned to left atrial rhythm; he remained in that rhythm when he was discharged.

** Case 2*

Si. Ha., a 13-year-old Indonesian boy, was referred to the clinic for the first time when he was 5 years old for the diagnosis and further management of cardiac abnormality. He was seen in the clinic very rarely because he lived in another city far away from Jakarta. He was the youngest of 6 children in the

family, and a product of normal pregnancy and uneventful delivery. His birth weight was 3.6 kg. The past history indicated that cyanosis was noticed by the parents when he was 3 months old, and persisted. His development was said to be poor, being able to sit by himself at 10 months and started walking at 18 months. There was history of recurrent respiratory infection, poor exercise tolerance and headache, but never suffered from dyspnea, cyanotic spells, nor squatting. In 1974 he had tonsillectomy.

Physical examination

Body weight was 28.5 kg., height 137 cm., a moderately build boy, central cyanosis, clubbing of the fingers and toes. Heart and pulse rate was 80/minute, respiration 26/minute, blood pressure 110/70 mmHg, no distention of jugular vein. The chest was asymmetric with bulging on the right side. Apex beat was seen and palpable on the right side of the chest. The heart was slightly enlarged, with increased right ventricular activity. The first heart sound was normal, the splitting of the second heart sound was rather wide with pulmonary component delayed and diminished. Ejection sound and ejection systolic murmur grade II-III/VI were heard in the 2nd-3rd Right Sternal Border; no thrill was felt. The lungs were clear on percussion and auscultation; the liver and spleen were not palpable; the pulse in the femoral arteries was of good quality. There were cyanosis and clubbing of the fingers and toes.

Laboratory data

Hemoglobin concentration was 18 gm.%, erythrocyte count 5.4 millions/cumm., hematocrit 53%, leucocyte count 6,400/cumm., and thrombocyte count 214,000/cumm. Chest X-ray and fluoroscopy revealed dextrocardia with situs inversus, right ventricular hypertrophy, and decreased lung vascularity. Phonocardiography was suggestive of valvular pulmonary stenosis. Vectocardiography revealed right ventricular hypertrophy, type C-

Electrocardiogram

1968 — QRS axis +20, QRS rate 100, P axis + 150 T axis + 135, PR interval 0.16", inverted P waves in I, II, aVL, upright P in III, aVR and aVF, tall peaked P in III, V2R, dominant R in V3 and V1R (= V2), dominant S in V2R — V6R. ECG diagnosis of mirror image dextrocardia, sinus rhythm, right atrial hypertrophy and right ventricular hypertrophy, was made.

In this patient, however, the left atrial rhythm did not persist for long. ECG recording on other occasions later on showed that the heart had returned to sinus rhythm.

Memodynamic studies

Catheterization and angiocardiography were done using no. 7 F catheter through the right v. saphena magna. Contrast material was injected in the right ventri-

cle. Frequent runs of atrial and ventricular extra systoles were encountered. Left ventricular angiography was not attempted. The result showed mirror image dextrocardia with tetralogy of fallot.

Discussion

Many authors have described various aspects of cyanotic congenital heart disease, including those of Double Outlet Right Ventricle and dextrocardia with Tetralogy of Fallot. Tausig and Bing (1949) reported a case of Double Outlet Right Ventricle which later was classified into 3 types by Witham (1957), and further into 2 groups by Neufeld et al. (1961). Mirowski et al. (1963) described ECG characteristics in patients with Double Outlet Right Ventricle (DORV) with pulmonary stenosis and its differentiation from Tetralogy of Fallot (TF). Sridaromont et al. (1976) in their review on DORV described 16 possible variations of DORV with regard to interrelations of great arteries and to location of the ventricular septal defect.

However, their association with the left atrial rhythm was hardly found in the literature. Mirowski et al. (1963) reported that 8 out of his series of 12 cases of left atrial rhythm were dextrocardia, while 4 were associated with normally placed heart. All patients were found to have concomitant heart defects, 4 cases were with TF all of which had Blalock-Taussig anastomosis, while only one in his series was with DORV.

It was mentioned that the left atrial rhythm may be found in a normal heart as well as in diseased ones.

The diagnosis of the left atrial rhythm in this study positively fulfilled the criteria proposed by various authors, such as Mirowski et al. (1963), or Constant (1973), Friedman (1971) and Chung (1971). It was reported by many authors abroad that patients with left atrial rhythm tended to suffer from frequent spontaneous or induced supraventricular arrhythmias. We also found the same situation in our 2 cases, where some difficulties were encountered during hemodynamic studies.

In case 1, the left atrial rhythm seemed to be persistent. But during observation, a transient spontaneous change from the left atrial rhythm into an unusual sinus rhythm with extreme left P

axis deviation was recorded. In this condition the P waves were inverted in II, III, aVR and aVL with normal PR interval. Chung (1971a) mentioned that as long as the P wave was inverted in lead aVR the origin of the P wave was most likely the sinus node, and he called this condition atypical sinus rhythm with extreme left P axis deviation. He related the left P axis deviation to the left atrial enlargement. This patient then spontaneously returned to the left atrial rhythm and remained that way when he was discharged.

In the second case the left atrial rhythm was probably transient. He came in sinus rhythm, and during observation the left atrial rhythm was recorded, but later spontaneously returned to sinus rhythm, and remained so when he was discharged.

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