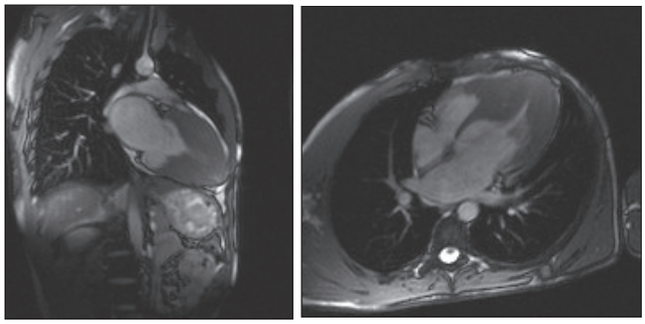

Figure 4 and 5. Transverse frontal cross- sectional magnetic resonance images. Normal basal and thickened apical segments

coronary artery disease and T waves normalized. In the other case, dilated cardiomyopathy developed and T waves regressed (2). Amplitude of giant T waves is far more correlated with the thickness of all myocardium than the thickness of sole apical myocardium (3). Horita at all. (4) reported that decreasing in the amplitude or disappearing T wave negativity is correlated with decreasing systolic function. In literature, some cases with hypertrophy restricted only by apical lateral, apical anterior or apical septum were reported. In these cases, spade- like ventriculogram do not occur (5).

Deep negative T waves may not be seen in all cases. T wave negativity may decrease or disappear with progression of disease. These changes may be early findings of other concomitant diseases (i.e coronary artery disease). On the other hand, as shown in our case, apical HCM may show normal T waves without concomitant diseases. As far as we explored, this is the first case report of Yamaguchi Syndrome without deep negative T waves and with findings of preexcitation in literature.

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A newborn with pentalogy of Cantrell and pulmonary hypoplasia

Bir yenidoğanda Cantrell pentalojisi ve pulmoner hipoplazi

To the Editors,

Ectopia cordis (EC) is rare, often fatal abnormality characterized by complete or partial displacement of heart out of thoracic cavity and occurs in 5.5-7.9 per 1 million live births (1). Four types of EC were defined: thoracic, abdominal, thoraco-abdominal and cervical (1, 2). If EC associated with deficiency of anterior diaphragm, midline supraumbilical abdominal wall defect, defect in diaphragmatic pericardium, various congenital intracardiac abnormalities, and defect of lower sternum, called pentalogy of Cantrell (PC) with an estimated incidence of 1/65.000- 1/200.000 live births (3). Chromosomal abnormalities, cleft lip-palate, pulmonary hypoplasia (PH), in some patients cranial, gastrointestinal, and renal abnormalities were reported in association (1, 2). We present a newborn with PC and PH.

Term female baby was born by vaginal delivery with irregular antenatal follow-up, with no history of drug abuse and consanguinity. She was transferred to our hospital two hours after birth because of extrathoracic heart. On the physical examination down slanted palpebral fissures, hypertelorism, micrognathia, short neck, extrathoracic heart with visible pulsations, supraumbilical abdominal wall defect, omphalocele between sternum and umbilicus were observed (Fig. 1). Echocardiography revealed four chambers; atrioventricular cavities on the left side were hypoplastic while atrioventricular cavities on the right side were dilated. There was single cardiac outlet [truncus arteriosus (TA)] originating from the ventricle on the right side, secundum atrial septal defect (ASD) and ventricular septal defect (VSD). No other congenital cranial, renal or gastrointestinal anomalies were present. Chromosomal analysis was 46, XX. She died on the 5th day before performing surgical procedure.

Autopsy findings; 6x4x2 cm heart mass with four chambers apex lying caudally was visible in front of the sternum. Left atrioventricular-atrioventricular valvular hypoplasia, ASD, VSD, TA originating from right ventricle, absent pericardium (naked heart), coarse trabeculae of right ventricle (L-loop position), four cusps of truncal valve were observed. Pulmonary veins meet inferior vena cava (VC) and inferior- superior VC drain to right atrium. Omphalocele was 5x5 cm, sternum was 3cm, xiphoid process was not present and both lungs were hypoplastic (2 lobes on the right, 3 lobes on the left with primitive upper lobe).

Toyama (4) made classification of PC. In class 1, cases have all five defects. Class 2 patients show four defects including intracardiac-

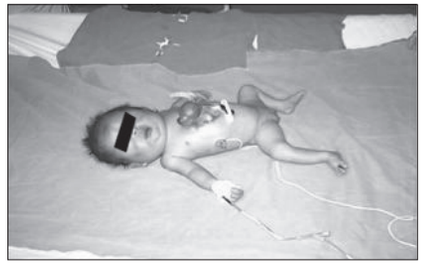

Figure 1. Dysmorphic face, ectopia cordis, lower sternal and supraumblical abdominal wall defects of the patient

Anadolu Kardiyol Derg
2009; 9: 518-21

ventral wall anomalies. Incomplete expression (class 3) is defined as various combination of abnormalities including sternal defect. Our patient was in class 1.

Antenatal diagnosis is important to organize neonatal, cardiovascular, and pediatric surgery team. Fetal-early postnatal cardiac and systemic evaluation for associated anomalies is necessary to determine surgery and assess prognosis. Diagnosis of our patient could be possible after delivery.

Life expectancy varies with site of ectopic heart and associated abnormalities (1-4). In a review, presence of intracardiac defects in EC may favor the prognosis that seemed to be poorer in complete PC and coexisting abnormalities (2). Correction of ventral hernia, diaphragmatic defects, and associated abnormalities is main treatment. Our patient died before surgery.

In conclusion, PC is rare anomaly and antenatal diagnosis is the most important point to be emphasized.

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Idiopathic twin aneurysm of right pulmonary artery diagnosed in a case 17 years after a successful surgical repair of ventricular septal defect

Başarılı ventriküler septal defekt onarımı gerçekleştirilmiş olguda 17 yıl sonra bulgulanan idiyopatik sağ pulmoner arter ikiz anevrizması

Pulmonary artery aneurysm is rarely seen in clinical practice (1). Aneurysm of pulmonary artery is relatively rare clinical finding, and appears mostly in association with significant cardiovascular or pulmonary abnormalities and is frequently caused by pulmonary hypertension (2). Different etiologies have been reviewed, but idiopathic lesions without other symptoms are seldom reported (3).

Our case was a 27-year-old male. He had undergone ventricular septal defect (VSD) repair in our clinic when he was 10 years old. Late period follow-up was event-free. He was referred to our clinic after the chest roentgenogram taken in another health facility due to upper respiratory tract infection. An abnormal shadow at right hilus region had been pointed out on the chest X-ray film (Fig. 1). On physical examination, the patient appeared generally well. Computed tomographic (CT) scan of the chest revealed a dilation of right pulmonary artery consistent with aneurysm at the level of hilus. Transthoracic echocardiography revealed no pathological finding and it was particularly reported that pulmonary arterial pressure and pulmonary valve were normal (Fig. 2). Thorax angiography with multislice CT of our case showed fusiform aneurysmal dilations of 18x20x38 mm at lobar branch and of 23x28x33 mm at middle lobar branch of right pulmonary artery (Fig. 3). With these findings, he was debated at our clinical council. Since he was asymptomatic and structure of pulmonary valve and pressure values were within normal limits, annual outpatient follow-up with thorax CT was recommended.

Aneurysm formation of the pulmonary artery has been reported to develop in patients due to several underlying etiologies. However, the natural history is not yet defined, and management remains controver-

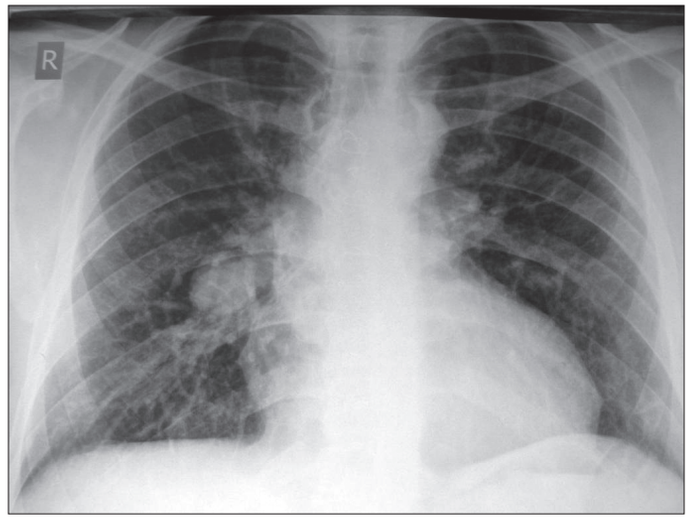

Figure 1. Chest X-ray of our case showing abnormal shadowing in the right pulmonary hilus

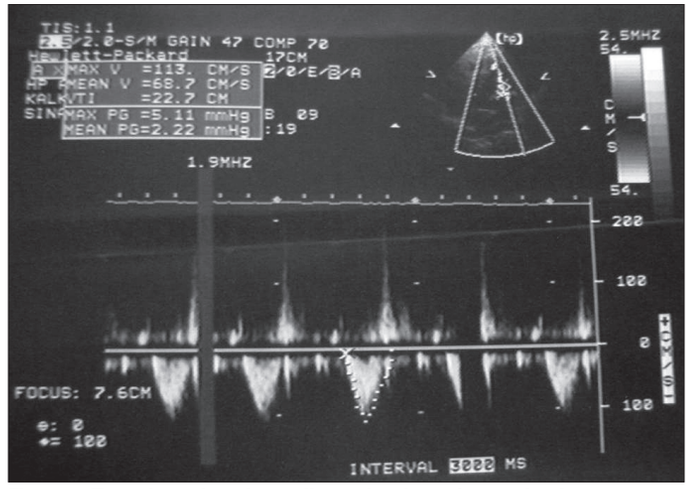

Figure 2. Transthoracic echocardiography view of normal pulmonary arterial pressure and structure of pulmonary valve