Total anomalous pulmonary venous connection (TAPVC) is a congenital cardiac anomaly that can mimic many cardiac and non-cardiac conditions during infancy and the neonatal period [1]. It is an emergency cardiac lesion that requires immediate surgical intervention. Echocardiography with color Doppler mapping can provide detailed information about cardiac pathology in infants with TAPVC. The need for more invasive procedures, such as cardiac catheterization, in infants and neonates with TAPVC must be justified based on the association of the disease with other intracardiac lesions or the need for documentation of pulmonary venous obstruction. This study assessed the accuracy of echocardiography in assessing sick infants and children with TAPVC.
Patients and Methods

From July 1, 1993, to December 31, 1999, TAPVC was diagnosed in 15 consecutive patients initially at a primary care center. The patients were transferred to the cardiovascular center of a tertiary medical hospital for surgical correction. Patients with TAPVC associated with heterotaxy syndrome were excluded from this study due to its frequent syndromic association with TAPVC. The clinical manifestations, plain chest roentgenograms, echocardiograms, magnetic resonance images, and surgical notes of these 15 patients were reviewed. We listed the most convincing echocardiographic features in all types of TAPVC. We compared echocardiographic features of coronary sinus TAPVC with those of the cor triatriatum for differentiation. All but one patient were less than 12 months old. Seconal (5–8 mg/kg/dose) administered per rectum was used for sedation. Suprasternal, parasternal, apical, and subcostal views were employed to explore the presence of TAPVC [1, 2]. Cardiac catheterization was performed in two patients to rule out mixed TAPVC. Magnetic resonance imaging was performed in one patient.

Fig. 1. Plain chest roentgenograms and magnetic resonance imaging study of total anomalous pulmonary venous connection (TAPVC). A) Cardiomegaly, prominent pulmonary trunk, and increased pulmonary vascular markings are seen in plain chest roentgenograms of patients with non-obstructive TAPVC. B) A snowman contour is visible on plain chest roentgenogram in a patient with supracardiac TAPVC. Four right-sided arrows denote the right superior vena cava, and four left-sided arrows indicate a vertical vein. C) A combination of normal heart size, passive pulmonary congestion, and a ground-glass appearance is seen on plain chest roentgenogram in a patient with obstructive infracardiac TAPVC. D) A descending vein (arrow) is shown penetrating the diaphragm and draining into the portal vein on magnetic resonance imaging study.
Results

Clinical profiles
The cardiac pathology of the 15 patients with TAPVC is shown in the Table. The left innominate vein, coronary sinus or right atrium, and portal vein were the sites of drainage of all cases of supracardiac, cardiac, and infracardiac TAPVC, respectively. Interatrial septal defect was found in all patients. Cardiomegaly, prominent pulmonary trunk, and increased pulmonary vascular markings were found on plain chest roentgenograms in 13 patients with non-obstructive TAPVC (Fig. 1A). A snowman contour could be seen in four patients with supracardiac TAPVC (Fig. 1B). A combination of normal heart size and passive pulmonary congestion, mimicking the condition of respiratory distress syndrome, was noted in two patients with infracardiac TAPVC with obstruction (Fig. 1C). Arterial blood gas showed severe hypoxemia (PaO₂ 25–35 mmHg) in two patients with obstructive infracardiac TAPVC. Oxygen saturation ranged from around 80% to 90% on oxymeter in 13 patients with non-obstructive supracardiac and cardiac TAPVC. Right atrial enlargement and right ventricular hypertrophy were found on electrocardiography and on frontal and lateral roentgenograms, in all patients except those with infracardiac TAPVC. One patient with cardiac TAPVC suffered from severe pulmonary hypertension, which was suprasystemic, and pulmonary vascular obstructive disease. He had a pulmonary-to-systemic resistance ratio of 0.9 due to a prolonged untreated course. Magnetic resonance imaging was employed in one patient to identify other associated intracardiac anomalies in addition to the infracardiac TAPVC to the portal vein (Fig. 1D).

Echocardiography
The suprasternal long axis and its rotational views are most useful for detecting supracardiac TAPVC (Fig. 2). Coronary sinus TAPVC can be diagnosed over the parasternal long-axis view (Fig. 3A), and its anterolateral tilting view. The only pitfall that can be encountered in echocardiography is cor triatriatum, which possesses a membrane much more vertical in the parasternal long-axis view and can be clearly differentiated from coronary sinus TAPVC by anterior tilting of the probe, during which this membrane remains in the left atrium receiving the pulmonary venous blood. With the aid of color Doppler, a turbulent jet through the ostium of the separating membrane can be visualized.

In patients with TAPVC to the coronary sinus, the pulmonary venous confluence and its drainage can be visualized over the apical four-chamber view (Fig. 3B), and its anteromedial tilting view, respectively (Fig. 3C). In case of cor triatriatum, a membrane partitioning the left atrium into two separate chambers can be seen horizontally in the apical four-chamber view. However, this horizontal membrane remains in the left atrium, in spite of the anterior tilting of the probe. A turbulent jet via the orifice of the limiting membrane can be visualized with the aid of color Doppler. In patients with

<table>
<thead>
<tr>
<th>Age</th>
<th>Sex</th>
<th>Type</th>
<th>Drainage</th>
<th>Flow profile</th>
<th>Associated lesion</th>
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<td>LINV</td>
<td>Phasic</td>
<td>ASD II</td>
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<tr>
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<tr>
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<td>CS</td>
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<td>PoV</td>
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<tr>
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<td>Infracardiac</td>
<td>PoV</td>
<td>Non-phasic</td>
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M = male; LINV = left innominate vein; ASD II = secundum atrial septal defect; PDA = patent ductus arteriosus; F = female; CS = coronary sinus; BSVC = bilateral superior vena cava; VSD = ventricular septal defect; RA = right atrium; CAVC = complete atrioventricular canal; SPMD = septum primum malposition defect; PVOD = pulmonary vascular obstructive disease; Pat = pulmonary atresia; PoV = portal vein; TOF = tetralogy of Fallot.
TAPVC draining directly to the right atrium, there were characteristic changes to the septum secundum and the septum primum over the apical four-chamber view (Fig. 4A). There was no evidence of pulmonary venous obstruction in patients with cardiac TAPVC.

In patients with TAPVC directly to the right atrium, septum primum malposition defect can be visualized over the subcostal transatrial view. In patients with infracardiac TAPVC, the course of drainage of the pulmonary venous confluence and its flow pattern can be easily detected over the subcostal transatrial view (Fig. 5).

**Discussion**

Thorough understanding of the development of the pulmonary venous system facilitates differentiation among TAPVCs in which there are persistent connections between the splanchnic venous plexus, cardinal venous system, and the pulmonary venous plexus [3–5]. TAPVC can mimic so many cardiac and noncardiac diseases with persistent pulmonary hypertension of the newborn that early differentiation and identification are essential before surgical intervention [6–9].

Clinically, patients with TAPVC may have congestive heart failure and/or profound cyanosis resulting from a balance between the interatrial communication and the patency of pulmonary venous flow. Adequacy of interatrial communication via the secundum atrial septal defect or septum primum malposition defect is suggestive of heart failure more than cyanosis. In this condition, anticongestive agents may have a better palliative effect than balloon atrial septostomy, which is important in the management of patients with TAPVC before transfer to a tertiary medical center for total correction. However, in cases with pulmonary venous obstruction at the atrial level due to a small and restrictive atrial septal communication, emergency balloon atrial septostomy is mandatory for survival before surgery. Precatheterization measurement of the size of the interatrial defect and evaluation of the flow pattern via the defect by echocardiography are informative for decision making [9].

On physical examination, a systolic ejection murmur and/or a loud pulmonary component of the second heart sound can be heard over the pulmonary area in patients with isolated TAPVC. Plain chest roentgenograms showed cardiomegaly and increased pulmonary vascular markings in patients with supracardiac and cardiac TAPVC. A snowman figure is found in patients with supracardiac TAPVC to the left innominate vein [6–8]. Ground-glass lung fields, which mimic those seen in respiratory distress syndrome in cyanotic premature infants, were noted in our two cases of infracardiac TAPVC with pulmonary venous obstruc-
Total Anomalous Pulmonary Venous Connection

Fig. 3. Echocardiographic images in a patient with total anomalous pulmonary venous connection to the coronary sinus. A) Parasternal long-axis view shows a dilated coronary sinus (CS) as a bulging horizontal membrane that slightly parallels the anteromedial wall of the left atrium. B) Apical four-chamber view shows the pulmonary venous confluence (star) that separates the left atrium laterally. C) View obtained by tilting anteromedially from the standard apical four-chamber view shows the pulmonary venous confluence (star), which can be traced to drain to the dilated CS. D) On the subcostal tilting or rotating view, the pulmonary venous confluence (star) can be traced to drain to the dilated CS.

In isolated TAPVC, there is a clear line of demarcation between the left atrium and the pulmonary venous confluence. Usually, a pulmonary venous confluence is situated posterior to the left atrium. Cor triatriatum can occasionally be mistaken for coronary sinus TAPVC on echocardiography if the section planes are not in true profile. Various tilting procedures of the standard precordial and subcostal views can be employed to differentiate these two anomalies.

Echocardiography can delineate the course and site of the anomalous pulmonary venous drainage as well as the obstruction of the pulmonary venous inflow from TAPVC by visualization of all (or some) of the pulmonary veins draining to the left atrium. In patients with isolated TAPVC, there is a clear line of demarcation between the left atrium and the pulmonary venous confluence. Usually, a pulmonary venous confluence is situated posterior to the left atrium. Cor triatriatum can occasionally be mistaken for coronary sinus TAPVC on echocardiography if the section planes are not in true profile. Various tilting procedures of the standard precordial and subcostal views can be employed to differentiate these two anomalies.

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subcostal short-axis rotation cut. Coronary sinus TAPVC can be detected and differentiated from cor triatriatum on the parasternal long-axis and apical four-chamber views and their tilt-axis views. Malalignment between the underdeveloped septum secundum and the leftward malpositioned septum primum, i.e., septum primum malposition defect, can contribute to the morphogenesis of TAPVC draining directly to the right atrium [18]. A non-phasic pattern of pulmonary venous flow can be detected by pulsed-wave Doppler in patients with congenital pulmonary vein stenosis [13, 14], and TAPVC with obstruction [15–17]. Pulsed-wave Doppler also shows the presence of acquired pulmonary venous obstruction following total correction for TAPVC [19].

In conclusion, various types of TAPVC can be accurately verified by echocardiography, except for mixed TAPVC, which was not found in this series. The adequacy of interatrial communication or obstruction at the atrial level in TAPVC should be assessed prior to
balloon atrial septostomy. A non-phasic pulmonary venous flow-characterized obstruction should be explored before and after surgery for TAPVC. Underdevelopment or absence of the septum secundum and malposition of the septum primum contribute to the morphogenesis of TAPVC directly to the right atrium.

References