Abnormal Eustachian Valve

GEN NIWAYAMA and VIBUL V. VADAKAN

Department of Pathology and Pediatrics, School of Medicine, University of California, Los Angeles, California, and Department of Pathology, Children's Hospital, State University of New York at Buffalo, U.S.A.

NIWAYAMA, G. and VADAKAN, V.V. *Abnormal Eustachian Valve*. Tohoku J. exp. Med., 1971, 104 (3), 263-287 — Congenital anomalies of the Eustachian valve were found in 3 patients. In a 12-year-old boy, the valve was abnormally enlarged and thickened, causing moderate constriction of the inferior vena cava orifice. A one-month-old infant exhibited aneurysmal dilatation in the midportion of the valve, combined with a right aortic arch and interventricular septal defect. A markedly enlarged, membranous Eustachian valve was present in a 2-month-old baby, with associated interventricular septal defect, transposition of the great vessels and bicuspid pulmonic valve. ——— congenital anomaly; Eustachian valve

Ever since interatrial septal defects have been included among congenital heart diseases which can be helped by surgical repair (Bailey *et al.* 1953, Gross *et al.* 1953, Bjork *et al.* 1954), exact knowledge of the normal anatomy of the right atrium and the recognition of deviations from the normal formation of its valves are no longer matters of mere embryologic and morphologic interest, but are rather emerging as actual clinical problems. The Eustachian and Thebesian valves, at the entrance of the inferior vena cava and of the coronary sinus, respectively, are normally both the remnants of the right valve of the sinus venosus. The existence of a valve at the opening of the inferior vena cava into the right atrium was first described by Bartolommeo Eustachius in 1563. The first case of pathologic persistence of this structure was reported by Lindes in 1865 in a heart which also exhibited a single atrial cavity and structures, later interpreted by Yater (1929) as persistent right and left venous valves. Minor abnormal persistence of the valves of the sinus venosus, resulting in a larger than usual Eustachian valve, Thebesian valve or Chiari's network, were reviewed and classified by Yater in 1929. Doucette and Knoblich (1963) recognized four anatomic patterns of pathological persistence of the right valve of the sinus venosus. Lucas and Schmidt (1968) mentioned 15 cases in the literature and broadened the classification offered by Doucette and Knoblich (1963). This paper represents an attempt to review the 70 cases which the authors have been able to trace in the literature, with additional report of three cases from the Children's Hospital of Buffalo.

Received for publication, December 22, 1970.

Reprint request should be sent to Dr. Niwayama, Department of Pathology, University of California, Los Angeles, California, 90024, U.S.A.

CASE 1

Maternal history: Unremarkable. Gestational period: full term, uncomplicated. Normal labor. Baby: vigorous and acyanotic. No resuscitation required. Birth weight: 8 pounds.

Medical history: Measles, German measles and pneumonia in early childhood. At the age of 7 years, a soft systolic heart murmur was detected by the family physician. However, this was not heard on subsequent examinations, according to the patient's mother. The patient remained in good physical health until 12 years of age. Two weeks prior to demise, he experienced an episode of cyanosis and light-headedness while wrestling. Because of the previous history of heart murmur, a medical clinic appointment was requested.

Physical examination: According to the residents note, there were no signs of symptoms characteristic of heart disease.

X-ray: Transverse diameter of chest: 28.6 cm, heart: 13.4 cm. Normal cardiac silhouette and lung fields.

Electrocardiogram: Severe left ventricular hypertrophy.

Clinical course: On the day following medical examination in the clinic, the child collapsed and died while running track at school.

Family history: Father died at age 35, with the clinical diagnosis of acute myocarditis. A younger sibling has "rheumatic heart disease." Two other siblings are well. Three paternal uncles died of undefined cardiac diseases at early ages.

Autopsy findings: Very well developed, well nourished white male. The bronchomediastinal lymph nodes were moderately enlarged and slightly congested. There was marked congestion of the neck veins, liver, spleen and kidneys. The lungs were well expanded with somewhat emphysematous appearance.

Significant necropsy findings were limited to the heart, which was markedly enlarged, measuring 9.7 cm × 11.5 cm. The left ventricle and papillary muscles were markedly hypertrophied, the ventricular wall measuring 13–19 mm, as compared with the right ventricle, 4–5 mm in thickness. The left atrium appeared slightly grayish-yellow. The right atrium was slightly dilated and the atrial wall slightly thickened. The Eustachian valve had a semilunar appearance and was abnormally large, measuring 2.4 cm at the margin, 2.1 cm in width and 2 mm in thickness. Its upper surface was smooth and regular. The undersurface revealed several fine band-like irregularities. The whole valve was firm, muscular and not fenestrated. The junction of the anterior inferior free margin with the septum was about 7 mm below the lower margin of the fossa ovalis and 1 cm above the commissure of the tricuspid cusps. The upper posterior junction of the valve was 9 mm above the upper margin of the fossa ovalis. Below the Eustachian valve, the coronary sinus and the Thebesian valve were normally located. The crista

Fig. 1.

Fig. 2.

Figs. 1 and 2. Superior view of the right atrium (case 1) E: Eustachian valve. SVC: Superior vena cava. T: tricuspid valve.

terminalis was prominent. The tuberculum Loweri was clearly recognizable.

The fibrous fossa ovalis measured 2.2 cm $\times$ 2 cm. The superior vena cava measured 4.2 cm in circumference at its entrance into the right atrium. The inferior vena cava was moderately dilated and measured 5.9 cm in circumference. The right atrium was partly divided by the anomalous Eustachian valve and the valve itself appeared to constrict the entrance of the inferior vena cava, causing marked congestion of the abdominal viscera. The heart valves were regular and measured: tricuspid 9.5 cm, pulmonic 6.9 cm, mitral 10.1 cm and aortic 5.9 cm in circumference.

Microscopic examination: Sections taken from the left ventricle revealed distinct hypertrophy of the cardiac musculature with microscopic appearance of severe chronic myocarditis. The finding was somewhat similar to that of an older stage of rheumatic lesions. Sections taken from the left atrium showed a moderate degree of endocardial fibrosis, which was considered secondary to severe myocardial damage to the left ventricle. Microscopic section taken from the Eustachian valve itself revealed normal cardiac muscle bundles which were continuous with the musculature of the right atrial wall.

The lungs showed marked congestion. There was slight to moderate mural thickening, especially of the media in small arterioles; spleen and kidneys were markedly congested.

Fig. 3. Microphotograph of the Eustachian valve. (H.E. stain)

CASE 2

Maternal history: Pregnancy complicated by elevated blood pressure and pedal edema. Spontaneous vaginal delivery complicated by prolapsed cord. No immediate neonatal complication.

Medical history: A transient heart murmur, of unspecified nature, was noted on the second day of life. The child was discharged without further diagnostic evaluation. He returned at 6 days of age with cyanosis, grade III/VI systolic heart murmur, heard equally over the entire precordium, and hepatomegaly, palpable one fingerbreadth below the right costal margin. During hospitalization bilateral staphylococcal conjunctivitis was noted, which precluded definitive evaluation of his cardiac status. He was readmitted at 3 weeks of age with a history of cyanosis, dyspnea, poor feeding and one apneic episode requiring resuscitation.

Physical examination: Pulse 120, respiration 32, lethargic. Mild cyanosis. HEENT: normal. Lungs: normal. Heart: PMI left of anterior axillary line. Grade IV/VI systolic murmur best heard at fourth intercostal space, left sternal border. No hepatosplenomegaly.

Laboratory data: Hemoglobin 13.3%. WBC 11,400. Urinalysis: within normal limits.

X-ray: Cardiothoracic ratio 5.8 cm/9.1 cm. Cardiac configuration unremarkable. Extensive pneumonic densities in upper halves of both lung fields and along right cardiac border.

Hospital course: The hospital stay was complicated by frequent cyanotic and apneic episodes requiring digitalization and use of antibiotics. Cardiac arrest occurred on the seventh day of hospitalization. Resuscitation was successful but endotracheal intubation with artificial ventilation was required. The child's condition deteriorated progressively and he expired at the age of 32 days.

At autopsy there were no significant findings in larynx, trachea and main bronchi. The pleural cavities were free of effusion. Both lungs were atelectatic. There was congestion of the neck veins, liver, spleen and kidneys.

The pericardial sac contained about 3 ml of slightly bloodtinged fluid. The heart was markedly enlarged. The right ventricle was markedly dilated and hypertrophied, up to 6-7 mm in thickness. The atria were unremarkable. The foramen ovale appeared as a slit-like opening. A small interventricular septal defect was present beneath the posterior cusp of the aortic valve, 4-5 mm in diameter. The valves measure: tricuspid 3.7 cm, pulmonic 2.6 cm, mitral 3.4 cm and aortic 2.2 cm in circumference. The coronary arteries appeared normal. The pulmonary artery was of normal caliber. The aorta showed a right-sided arch. The ductus arteriosus was closed. The superior vena cava was normal. The inferior vena cava revealed an abnormally elongated Eustachian valve. This thin, membranous structure exhibited an area of aneurysmal dilatation in its mid-

Fig. 4.

Fig. 5.

portion. The coronary sinus and Thebesian valve were normal.

A small hemangioma was present on the septal cusp of the tricuspid valve.

Microscopic examination: Non-contributory.

CASE 3

The third patient is a male infant with multiple congenital cardiac anomalies who died at 9 months of age from thromboembolism to the left internal carotid artery.

Autopsy findings: Larynx, trachea, main bronchi and lungs were not remarkable except for small area of atelectasis in basal segment of left lower lobe. Pleural cavities were free of effusion.

The pericardial sac contained a small amount of clear amber fluid. The heart was enlarged, 5.7 cm in vertical and 5.6 cm in horizontal plane. The right ventricle was markedly dilated and hypertrophied, 7–9 mm in thickness, as against 3–6 mm of the left ventricular wall. The atria were not remarkable. A minute opening was present at the foramen ovale. There was a large interventricular septal defect, measuring 2 cm vertically and 1.3 cm horizontally in diameter. The upper edge of the defect was located 12 mm below the pulmonary valve. The tricuspid valve measured 4.5 cm, mitral valve 4.2 cm. The chordae tendineae of the anterior cusp of the mitral valve attached themselves to an area just beneath the crista supraventricularis of the right ventricle. Some of the chordae tendineae of the posterior cusp of the mitral valve showed similar adhesions to the right side of the right ventricular wall. The aorta arose from the right ventricle and measured 3.9 cm in circumference. Three aortic cusps and two coronary ostia were noted. Petechiae were seen on the adventitial layer of the ascending portion of the aorta. The pulmonary ostium with its two cusps arose from the left ventricle and measured 2.4 cm in circumference. Probe patency was present of the ductus arteriosus. The left subclavian artery took its origin at a point proximal to the ductus arteriosus. The superior and inferior venae cavae emptied normally into the right atrium. The coronary sinus and Thebesian valve were unremarkable. Small recent thrombi were present at the lower margin of the interventricular septal defect and also on the posterior surface of the anterior cusp of the mitral valve.

The Eustachian valve was abnormally enlarged, measuring 11 mm at its greatest width. It was thin and membranous except for a small muscular portion on the side facing the interatrial septum.

Figs. 4 and 5. Right atrium and ventricle (Case 2). C: Coronary sinus. E: Eustachian valve. IVC: Inferior vena cava. SVC: Superior vena cava. T: Tricuspid valve. A dot between C and T indicates a small hemangioma of the tricuspid valve.

Fig. 6. Anterior view of the right atrium and ventricle. (Case 3)

Fig 7. The "a" is a diagram of Fig. 6 (Case 3). The "b" is a diagram of the reconstructed interior view of the right atrium. C: coronary sinus. E: Eustachian valve. SVC: Superior vena cava. T: Tricuspid valve.

DISCUSSION

During the embryonic development of the heart, the single atrial cavity is divided into two definite chambers in the following manner: the septum primum arises from the mid-dorsal wall of the atrium and progressed downward toward the

atrioventricular opening. Two swellings appear in the wall of the atrioventricular canal, forming the septum intermedium. The narrow opening between the septum primum and the septum intermedium is the ostium primum. Before the continued growth of the septum primum completely closes the ostium primum, an area of the septum near its cephalic attachment to the atrial wall thins out and breaks through, forming the ostium secundum. In the right atrium, a new partition grows downward from the cephalic and ventral part of the atrial wall as the septum secundum. Its downward growth ceases before it completely covers the new opening of the septum primum, leaving a communication known as the foramen ovale (Henry Gray 1959)

The right atrium of the adult heart is developed from the sinus venosus, the primitive atrium and the right half of the atrial canal. The sinus venosus opens into the dorsal wall of the primitive atrium through the vertically placed sinoatrial foramen. This slit-like opening is guarded by two valves, the right and left valvulae venosae. Above the opening, the two venous valves unite into a single fold, forming the septum spurium. Below, these valves also fuse and are continued toward the atrial canal (Hickie 1956). The muscular fused portion contracts during atrial systole, thus preventing backflow of atrial blood into the sinus venosus (Lucas and Schmidt 1968). As development progresses, the orifice of the transverse part of the sinus venosus is drawn into the posterior wall of the right atrium, and the left valve and septum spurium fuse with the septum secundum. The upper part of the sinus becomes the opening for the superior vena cava. The inferior part is divided by the further growth of the right venous valve into two openings, the inferior vena cava and the coronary sinus.

Bands of muscle form between the openings of the coronary sinus, the superior and inferior venae cavae, so that the original sinus opening and right sinus valve are divided into three parts: (1) the superior caval opening ridged above and laterally by the remains of the upper part of the right sinus valve—the crista terminalis; (2) the inferior caval opening bounded in front by the middle half of the right venous valve—the Eustachian valve; and (3) the coronary sinus bounded on the right by the anterior and of the right sinus valve—the Thebesian valve (Hickie 1956).

The right horn is absorbed into the right atrium, forming the sinus venarum. The primitive atrium becomes the auricular appendage and the area of the atrium provided with muscoli pectinati. The right half of the atrial canal forms the smooth area above the atrioventricular orifice.

In the fetal heart, the free concave cranial border of the Eustachian valve is directed upward and medially, channeling the blood stream from the inferior vena cava through the foramen ovale into the left atrium (Yater 1929).

The adult Eustachian valve is a musculomembranous semilunar fold, attached to the ventral and left margin of the orifice of the inferior vena cava. Its concave free margin ends in two cornua, the left of which is continuous with the ventral margin of the limbus fossae ovalis and the right spreads out on the atrial wall.

Author	Year	Age	Sex	Clinical data
Lindes	1865	4 hrs	M	Born during eighth month of gestation Ascites noted at birth
Maier	1867	33 yrs	F	Chronic bronchitis
Von Rokitansky	1875			
Lauenstein (5 cases)	1876			Two female and three male patients
Moore (also mentions two similar cases)	1883	33 yrs	F	Tuberculosis
Leo	1886	2 yrs	F	Intense cyanosis since birth
Preisz	1890	Stillbirth	M	
Stadler	1890	3 mo.	F	
Przewoski	1897			
Chiari	1897	24 yrs	M	Pulmonary embolism from thrombus in network
Chiari	1897	43 yrs	M	Chronic disseminated tuberculosis
Chiari	1897	30 yrs	M	Aortic valvular endocarditis
Chiari	1897	45 yrs	F	Chronic pulmonary tuberculosis

Cause of death	Eustachian valve anomaly	Associated cardiac defects
	Persistent right and left venous valves	Single atrial cavity Common atrioventricular canal Stenosis of pulmonary valve Patent ductus arteriosus Persistent left SVC
"Hydrothorax" "Hydropericardium" "Hydrops Ascites"	Two folds in right atrium representing persistent right venous valve and remnant of septum primum	Dilatation and hypertrophy of right atrium and ventricle Mitral stenosis Left atrial dilatation Hypoplastic left ventricle
	Persistent right venous valve	
	Eustachian and Thebesian valves formed by one extensive membrane Simple membrane in one case, fenestrated membrane in three cases, network of fine fibers in fifth case	Patent foramen ovale in one case
Tuberculosis	Large muscular fold beginning below fossa ovalis and stretching from interatrial septum near of right atrium, representing exaggerated Eustachian valve with excessive development of inferior sinus septum	
	Sail-like Eustachian valve with several perforations	Absence of tricuspid valve Atresia of pulmonic valve Hypoplasia of right ventricle Patent foramen ovale Patent ductus arteriosus
	Sieve-like flap at right border of IVC and coronary sinus corresponding to right venous valve	
	Fold arising from IVC representing remnant of right venous valve	Stenotic bicuspid pulmonic valve Ventricular septal defect Two pulmonary vein orifices in left atrium (Cheilognathopalato-schisis)
	Chiari's network containing thrombus	
Pulmonary embolism	Chiari's network	
Tuberculosis	Chiari's network	Aortic valvular endocarditis
Aortic valvular endocarditis	Chiari's network	
Tuberculosis	Chiari's network	Mitral valve stenosis

Author	Year	Age	Sex	Clinical data
Chiari	1897	43 yrs	F	Cardiac disease
Chiari	1897	40 yrs	F	Chronic disseminated tuberculosis
Chiari	1897			Museum preparation
Chiari	1897	35 yrs	F	Cardiac disease
Chiari	1897	17 d	M	Acute "Enterokatarrh"
Chiari	1897	30 yrs	M	Chronic pulmonary tuberculosis
Chiari	1897	30 yrs	M	
A. Weber	1898			
LeCount	1901			
Looser	1902			Thrombi in both femoral veins Hemorrhagic embolic infarct in left lung
Ebbinghaus	1904	52 yrs	F	Frequent epistaxes Apoplexies at ages 38 and 40 with right hemiplegia
Wortmann	1909	12 d	M	
Thilo (5 cases)	1909			
Von Moellendorff	1912			

Cause of death	Eustachian valve anomaly	Associated cardiac defects
		Left atrial dilatation Hypertrophy and dilatation of right atrium and ventricle
Cardiac failure	Chiari's network	
Tuberculosis	Chiari's network	
	Chiari's network	
Cardiac failure	Chiari's network	Tricuspid valvular endocarditis Hypertrophy of right ventricle Mitral valvular stenosis
Same	Chiari's network	
Same	Chiari's network	Patent foramen ovale
Cardiac failure	Chiari's network	Mitral valvular endocarditis with stenosis Aortic valvular endocarditis with insufficiency
	Chiari's network derived from right venous valve and inferior sinus septum, with smaller network representing remnants of left venous valve	
	Chiari's network entirely below orifice of inferior vena cava	
	Chiari's network containing thrombus, considered remnants of right venous valve, left venous valve, septum spurium and sinus septum	
Hemorrhage into right basal ganglia of brain	Chiari's network	Membranous interatrial septum with eleven foramina Persistent ostium primum Persistent ostium secundum Hypertrophy of right ventricle
	Bulging, sac-like membrane containing thrombus considered residuum of right venous valve, with an independent network along orifice of IVC representing remnant of left venous valve	Tricuspid stenosis Stenotic right ventricular outflow tract Transposition of great vessels
	Reticula resembling Chiari's networks in right atrium. One case contains thrombus in network	
	Remnants of both right and left venous valves present. Sinus septum had not divided right venous valve into Eustachian and Thebesian valves.	

Author	Year	Age	Sex	Clinical data
Sternberg	1913	21 yrs	F	Aortic and mitral endocarditis following tonsillitis
Haas	1916			Thrombus in left femoral vein, embolus in network
F.P. Weber	1920	25 yrs	M	"Sarcomatous tumor" of left kidney, metastatic
F.P. Weber	1920	29 yrs	M	Exertional dyspnea since age 11 years Systolic murmur Pulmonary tuberculosis
Jordan	1926	65 yrs	M	Carcinoma of prostate
Jordan	1926	71 yrs	M	Kidney disease
Wurm	1927	3 mo.	M	Race: Anamese Cyanosis, mild Multiple episodes of dyspnea and cyanosis preceding death
Yater (3 cases)	1929			
Yater (2 cases)	1929			
Helwig	1932	67 yrs	F	Dyspnea and auricular fibrillation
Helwig	1932	43 yrs	F	Rheumatic fever during childhood Thrombus in left femoral artery→A-K amputation
Helwig	1932	38 yrs	F	Exertional dyspnea x 10 yrs Auricular fibrillation Right hemiplegia
Helwig	1932	50 yrs	M	Pain in left chest and left arm x 5 years
Helwig	1932	21 yrs	M	Spindle cell sarcoma, right thigh
Helwig	1932	24 yrs	M	Spongioblastoma of cerebral peduncle
Helwig	1932	53 yrs	F	Anemia and spinal cord degeneration

Cause of death	Eustachian valve anomaly	Associated cardiac defects
Same	Abnormally enlarged Eustachian valve	Patent foramen ovale Mitral valvular endocarditis Aortic valvular endocarditis
	Three groups of threads forming a network replacing the Eustachian and Thebesian valves. Embolus, 6 cm long, ensnared by network	
Same	Chiari's network	
Tuberculosis	Chiari's network	Dilatation and hypertrophy of right atrium Thrombus attached to chordae tendineae of tricuspid valve Contracted pulmonary orifice with thickened semilunar cusps. Adherent thrombi in pulmonary artery
Same	Chiari's network	
Same	Chiari's network	
	Chiari's network	Corrected transposition of great vessels Coarctation of aorta Tricuspid insufficiency Patent foramen ovale
	Chiari's network	
	Both Eustachian and Thebesian valves formed of one membrane	
Cardiac failure	Chiari's network	Mitral valvular stenosis and regurgitation Dilated left atrium (capacity 1000 cc) (Atrophic left lung, diameter 5 cm)
Cardiac failure	Chiari's network	Mitral stenosis Tricuspid stenosis Bilateral atrial dilatation Dilated coronary sinus
Cardiac failure	Chiari's network	Mitral stenosis
Heart failure Pulmonary edema	Chiari's network	Myocardial infarct, anterior 2/3 of interventricular septum
Same	Chiari's network	
Same	Chiari's network	
Same	Chiari's network	

Author	Year	Age	Sex	Clinical data
Helwig	1932			Museum preparation
Gomberg	1933	42 yrs	F	Postpartum pulmonary embolism
Kettler	1934	60 yrs	M	Pulmonary embolism
Ruggieri	1935	42 yrs	F	Cyanosis, systolic murmur since age 12 years Syphilis in adult life
Ponhold	1937	2 d	F	Cyanosis noted on second day of life
Dubin and Hollinshead	1944	5 min	F	Race: Negro; Heart failure
Kjellberg	1955	6 yrs	M	Pulmonic stenosis
Hickie	1956	47 yrs	M	Progressive exertional dyspnea Recurrent chest infections Left precordial pain Paroxysmal nocturnal dyspnea
Hickie	1956	27 yas	M	Progressive dyspnea; recurrent bronchitis
Rossall and Caldwell	1957	19 yrs	M	Varicose veins, hepatomegaly, pleural effusions, ascites, jaundice

Cause of death	Eustachian valve anomaly	Associated cardiac defects
	Chiari's network	
Same	Abnormally enlarged Eustachian valve	Defect of foramen I Hypoplasia of right ventricle Small pulmonary trunk
Same	Abnormally enlarged Eustachian valve	Patent foramen ovale "Wide" pulmonary trunk
Heart failure	Abnormally enlarged Eustachian valve Microscopic: Eustachian valve consists of dense connective tissue containing many thick elastic fibers	Patent foramen ovale "Narrow" pulmonary trunk
Heart failure	Abnormally enlarged Eustachian valve	Patent foramen ovale Patent ductus arteriosus Hypoplasia of tricuspid valve Hypoplasia of right ventricle Complete subvalvular pulmonic atresia Membranous interventricular septal defect
Heart failure	2x3 cm large membranous septum with few fenestrations at edges, attached superiorly to crista terminalis, inferiorly to tricuspid valve ring Microscopic: Dense connective tissue with bundles of elastic fibers and cardiac muscle. Each surface of Eustachian valve is covered by single layer of endothelium	Patent foramen ovale Patent ductus arteriosus Bicuspid hypoplastic tricuspid valve Hypoplasia of pulmonary trunk
Heart failure	Abnormally enlarged Eustachian valve	Infundibular pulmonic stenosis, surgically corrected
Died on operating table during atrio-septopexy	Eustachian valve divides right atrium into two chambers, the sinus venarum and the dilated right auricular appendage. Valve is perforated, 6.2 cm along free edge, 2.1 cm wide, 0.5 cm thick, with Chiari's network arising from right edge	Patent foramen ovale Atrial septal defect Tricuspid valvular insufficiency Mitral valvular stenosis Dilated right atrium Dilatation and hypertrophy of right ventricle
Died postoperatively following atrio-septopexy	Muscular Eustachian valve, 4.7 cm along free edge, 1.5 cm wide, 0.7 cm thick	Atrial septal defect Dilatation of right atrium Dilatation and hypertrophy of right ventricle Anomalous pulmonary venous return, partial, into right SVC Dilated pulmonary artery
Cardiac cirrhosis	Thin fibrous diaphragm-like Eustachian valve almost completely obstructing orifice of IVC, 4.4 cm along free border, 2.6 cm in depth.	

Author	Year	Age	Sex	Clinical data
Scholmerich	1962			
Doucette and Knoblich	1963	6 wks	M	
Kauffman and Andersen	1963	2 d	M	Cyanosis and respiratory distress starting at 12 hrs of age
Black, Smith and Goodale	1964	30 yrs	F	Cyanosis since birth Heart murmur since age 12
Case 1	1970	12 yrs	M	Transient systolic heart murmur heard at age 7 yrs. One episode of cyanosis and light-headedness two weeks before death
Case 2	1970	1 mo	M	Transient heart murmur noted on second day of life, followed by marked cyanosis, loud systolic murmur, hepatomegaly, apneic episodes
Case 3	1970	9 mo	M	

Cause of death	Eustachian valve anomaly	Associated cardiac defects
	Microscopic: Surface lined by single layer of flattened endothelium, moderately cellular connective tissue intermingled with numerous small elastic fibers. No cardiac musculature within valve itself. Small arterioles and dilated venules present within substance of valve.	
	Abnormally enlarged Eustachian valve excluding IVC and coronary sinus from right atrium while SVC drained normally.	
Postoperative death following modified Pott's procedure	Right atrium completely divided by muscular septum showing three perforations. It straddled the medial portion of the tricuspid valve and lay to the right of the SVC, IVC and coronary sinus. It separated atrial appendage from the sinus the venarum and terminated ventrocephalad in the right wall of the SVC.	Patent foramen ovale Patent ductus arteriosus Hypoplasia of tricuspid valve Atresia of pulmonary valve Hypoplasia of right ventricle
	Persistent venous valves	Hypoplasia of tricuspid valve Pulmonary atresia Multiple coronary artery-ventricular communications Focal necrosis, right ventricular myocardium
Postoperative infection	Failure of regression of right valve of sinus venosus, causing tunneling of IVC between the two leaves of the atrial septum into left atrium	(Multiple pulmonary arterio-venous fistulae)
Sudden death during exercise	Semilunar Eustachian valve, 2.4 cm at margin, 2.1 cm in width and 2 mm in thickness, constricting IVC orifice. Microscopic: Normal cardiac muscle bundles continuous with musculature of right atrial wall	Left ventricular hypertrophy Microscopic appearance of severe chronic myocarditis in sections from left ventricle
Cardiac failure	Thin, membranous, abnormally elongated Eustachian valve exhibiting an area of aneurysmal dilatation in its mid-portion	Dilatation and hypertrophy of right ventricle Patent foramen ovale Interventricular septal defect Right-sided aortic arch Hemangioma, septal cusp of tricuspid valve
Thromboembolism in left internal carotid artery	Thin, membranous, abnormally enlarged Eustachian valve, 11 mm at greatest width	Dilatation and hypertrophy of right ventricle Patent foramen ovale Interventricular septal defect Transposition of great vessels Patent ductus arteriosus

Its adherent border is a transverse muscular ridge continuous with the anterior margin of the inferior caval orifice (Henry Gray 1959). One of its surfaces is turned laterally toward the atrium and the other medially toward the inferior vena cava. The valve in the adult is usually rudimentary and has little, if any, functional significance. It is variable in size with an average length at its free edge of 3.4 cm and average width of 0.35 cm. It may be a single fold or fenestrated or may consist of a network of threads or even be entirely absent (Yater 1929).

In the animal kingdom, the venous valves may persist partly or entirely, as shown by Born (1888) in mammals and by Rose (1890) in monotremes and marsupials. Weber (1904) quoted Grosser as having found a persistence of the left venous valve in a certain number of Cheiroptera, and Devez in the American Didelphys. The Echidna (monotreme) has a valve for each vena cava and a valve common to both. The Ornithorynchus has a common internal valve represented by a simple musculomembranous ridge. Other animals having an internal valve in the inferior vena cava are the Myrmecopha tetradaetyla, the jaguar and the cabai.

In man, the Eustachian valve was first described in 1563 by Bartolomeno Eustachius (1563). Its existence was denied by Bauhin in 1605. Harvey, in his work (*Exercitatio Anatomica de Motu Cordis et Sanguinis in Animalibus* published in 1628), does not mention the Eustachian valve. Winslow gave the first accurate description in 1717, and was also the first person to suggest its fetal function. Sabatier (1774) and Barclay *et al.* (1944) later established that the valve directed blood across the foramen ovale into the left atrium in fetal life.

Anomalies of the Eustachian valve consist of variations in size, extent, muscularity, and formation of Chiari's network (Lindes 1865, Maier 1867, Von Rokitsansky 1875, Lauenstein 1876, Moore 1883, Leo 1886, Preisz 1890, Stadler 1890, Przewoski 1896, Chiari 1897, Le Count 1901, Loose 1902, Ebbinghaus 1904, Thilo 1909, Wormann 1909, Von Moellendorff 1912, Sternberg 1913, Haas 1916, Weber 1920, Jordan 1926, Wurm 1927, Yater 1929, Helwig 1932, Gomberg 1933, Kettler 1934, Ruggieri 1935, Ponhold 1937, Dubin and Hollinshead 1944, Gross *et al.* 1953, Kjellberg 1955, Hickie 1956, Rossall and Caldwell 1957, Scholmerich *et al.* 1962, Doucette and Knoblich 1963, Kauffman and Andersen 1963, Black *et al.* 1964, Lucas and Schmidt 1968).

CLASSIFICATIONS

Minor abnormal persistence of the valves of the sinus venosus, resulting in larger than usual Eustachian valves, were reviewed and classified by Yater in 1929. In his study comprising 120 adult hearts, all the Eustachian valves conformed to one or another of six anatomic types. A brief description of each type follows:

- 1) Absence of Eustachian valve (17 cases). The sinus septum and anterior rim of the inferior vena cava were all that denoted the previous existence of the right venous valve, except for the Thebesian valve, which was present in 16 cases

and absent in one case.

2) The Eustachian valve present as a simple non-fenestrated flap or membrane (69 cases). The usual form was crescentic. It varied from a narrow little fold along the anterior edge of the orifice of the inferior vena cava, to a broad membrane 2.3 cm wide. The valve in some cases was a thin, flabby, transparent membrane distinct from the rim of the inferior vena cava. In others, it was a taut fold merging with the rim of the inferior vena cava and the wall of the right atrium. It sometimes contained muscle fibers, especially near the attached edge.

3) The Eustachian valve present as a fenestrated semilunar membrane (22 cases).

4) The Eustachian valve formed in part or completely of a network of threads (7 cases).

5) Both the Eustachian and Thebesian valves formed of a single membrane (2 cases).

6) Chiari's network (3 cases). Usually Chiari's networks are persistent right valves and extend from the crista terminalis or tuberculum Loweri to the Eustachian and/or Thebesian valves.

In these cases of Yater (1929), the average width was 0.35 cm and the average length at the free edge was 3.42 cm. The minimal width was 0.15 cm and maximal width 2.3 cm. The minimal length at the free edge was 1.7 cm and maximal length 7.5 cm.

The lesions previously described represent minor abnormal variation or persistence of the right venous valve of the sinus venosus which are usually of little or no hemodynamic consequence. Pathologic persistence of the right valve of the sinus venosus, producing subdivision of the right atrium and on rare occasions causing obstruction to systemic venous return, is an uncommon condition. In addition to our cases, we have been able to trace in the literature only 70 other cases.

Six anatomic patterns are recognized by Lucas and Schmidt (1968):

1) In the complete form, the right atrium is divided into two chambers, the sinus venarum and the right atrium proper, by a membranous right valve which extends from the crista terminalis laterally to the medial wall, and from the superior vena cava above to a point below the coronary sinus. The sinus venarum receives the inferior vena cava, the superior vena cava and the coronary sinus. The true right atrium has the right auricular appendage and communicates with the right ventricle by way of the tricupsid valve. Variable-sized defects in the abnormal valve allow communications between the sinus venarum and the true right atrium. The foramen ovale also allows communication from the sinus venarum to the left atrium.

2) The superior and inferior venae cavae empty into the sinus venarum while the coronary sinus empties normally into the right atrium proper.

3) Only the inferior vena cava is within the sinus venarum. The superior

vena cava and the coronary sinus drain normally into the true right atrium. The persistent valve fuses with the septum secundum at the foramen ovale and blood from the inferior vena cava drains through this opening into the left atrium.

4) Only the ostium of the coronary sinus is excluded. The superior and inferior venae cavae drain into the true right atrium.

5) The inferior vena cava and the coronary sinus are both excluded from the right atrium while the superior vena cava drains normally.

6) The persistent right valve is not within the right atrium but at the junction of the inferior vena cava and the right atrium, causing partial or complete obstruction of the inferior vena cava.

The first patient presented in this report has an Eustachian valve anomaly which appears to belong to the third anatomic pattern listed above.

Most of the cases previously reported exhibited some form of associated right heart anomaly, usually in the form of pulmonary atresia, hypoplastic tricuspid valve and right ventricle. Significant inferior vena cava obstruction has been previously reported in only one patient (Rossall and Caldwell 1957). In our case, it was most likely responsible for his sudden demise. This obstruction to systemic venous return, evidenced by marked degree of congestion of the neck veins, liver, spleen, kidneys and other intraabdominal organs, was of acute nature. The young adult reported by Rossall and Caldwell (1957) had chronic obstruction and had as a consequence developed collateral venous pathways and centrolobular necrosis of the liver prior to his eventual demise.

The second patient presents, among other congenital defects, an abnormality of the Eustachian valve which had been previously reported on only one occasion by Wortmann (1909), in an infant with a bulging, sac-like membrane containing a thrombus in place of an Eustachian valve. This particular aneurysm, present in our second patient, was probably of no hemodynamic importance. The associated interventricular septal defect was undoubtedly the primary factor causing heart failure and death. This abnormal valve again separated the inferior vena cava from the superior vena cava and the coronary sinus and would belong to the third anatomic pattern in the classification of Lucas and Schmidt (1968).

The third patient had, among numerous other congenital cardiac defects, an abnormally enlarged Eustachian valve. The right atrium was partitioned into the sinus venarum containing the inferior vena cava entrance, and the right atrium proper, containing the orifices of the superior vena cava and the coronary sinus. Again, such an arrangement falls into the third anatomic type in the classification of Lucas and Schmidt (1968). As in the second case, the Eustachian valve anomaly is not believed to have been hemodynamically significant.

In view of the rapidly increasing number of intracardiac operations performed nowadays, the authors wish to emphasize the importance to be aware of the structural anomaly of the Eustachian valve. It can be a surgically correctable cause of heart failure. It can also confuse the surgeon who tries to manually delineate the anatomy of the right atrium, as happened in the two cases

reported by Hickie (1956).

SUMMARY

Three cases of congenital cardiac anomalies involving the Eustachian valve are presented. The first is a case of a 12-year-old boy with findings of severe chronic myocarditis combined with an anomalous Eustachian valve which divided the right atrium and caused moderate constriction of the inferior vena cava orifice, resulting in marked congestion of the intraabdominal viscera. The second case, a 30-day-old male infant, exhibited an area of aneurysmal dilatation in the midportion of the Eustachian valve associated with interventricular septal defect and right aortic arch. The third case, a 9-month-old male child with interventricular septal defect, transposition of the great vessels, bicuspid pulmonic valve and anomalous origin of the left subclavian artery, had an abnormally enlarged, membranous Eustachian valve. A review of similar anomalies reported in the literature and of the classification of minor variations and major anomalies is presented.

Acknowledgment

We wish to express our gratitude to Dr. Kornel L. Terplan for his suggestions in the preparation of this paper, and to Mr. T. Llyle Keith for his technical assistance.

References

- 1) Bailey, C.P., Bolton, H.E., Jamison, W.L. & Neptune, W.B. Atrio-septo-pxy for interatrial septal defects. *J. thorac. Surg.*, 1953, **26**, 184-219.
- 2) Barclay, A., Franklin, J. & Prichard, M. *The Foetal Circulation*, Blackwell, London, 1944.
- 3) Bauhin, C. *Theatrum anatomica*. (1605) (cited by Hickie in 18).
- 4) Bjork, V.O., Crafoord, C., Jonsson, B., Kjellberg, S.R. & Rudhe, U. Atrial septal defects. *Acta. chir. scand.*, 1954, **107**, 499-515.
- 5) Born, G. Ueber der Bildung der Klappen, Ostien und Schweidewaende im Saeugetierherzen. *Anat. Anz.*, 1888, **3**, 606-612.
- 6) Black, H., Smith, G.T. & Goodale, W.T. Anomalous inferior vena cava draining into the left atrium associated with intact interatrial septum and multiple pulmonary arteriovenous fistulae. *Circulation*, 1964, **29**, 258-267.
- 7) Chiari, H. Ueber Netzbildungen im rechten Vorhofe des Herzens. *Beitr. path. Anat.* 1897, **22**, 1-10.
- 8) Doucette, J. & Knoblich, R. Persistent right valve of the sinus venosus. *Arch. Path.*, 1963, **75**, 105-112.
- 9) Dubin, I.N. & Hollinshead, W.H. Congenitally insufficient tricuspid valve accompanied by an anomalous septum in the right atrium. *Arch. Path.*, 1944, **38**, 225-228.
- 10) Ebbinghaus, H. Zur Kasuistik der kongenitalen Herzfehler und deren möglichen Folgen. *Münch. med. Wschr.*, 1904, **51**, 797. (cited by Yater in 49).
- 11) Eustachius, B. *Opuscula. Anatomica*, 1563 (cited by Hickie in 18).
- 12) Gombert, H. Beiträge zur Pathologie der Vorhofsschiedewand des Herzens. *Beitr. Path. Anat.*, 1933, **91**, 483-502.
- 13) Gross, R.E., Watkins, E., Jr., Pomeranz, A.A. & Goldsmith, E.I. A method for surgical closure of interauricular septal defects. *Surg. Gynec. Obstet.*, 1953, **96**, 1-23.

- 14) Haas, W. Über einen weiteren Fall von Netzbildungen im rechten Vorhof mit einem in denselben verfangenen Embolus. Inaug. Dis. Karlsruhe. 1916 (cited by Yater in 49).
- 15) Harvey, W. *Exercitatio Anatomica de Motu Cordis et Sanguinis in Animalibus*. (1628) (cited by Hickie in 18).
- 16) Helwig, F.C. The frequency of anomalous reticula in the right atrium of the human heart "Chiari network": report of 8 cases. *Amer. J. Path.*, 1932, 8, 73-79.
- 17) Henry Gray, F.R.S. *Anatomy of the Human Body*. 27th edition. Lea & Febiger, Philadelphia, 1959, pp. 567-601.
- 18) Hickie, J.B. The valve of the inferior vena cava. *Brit. Heart J.*, 1956, 18, 320-326.
- 19) Jordan, W.R. Two cases of Chiari's network. *Arch. Path.*, 1926, 2, 840-843.
- 20) Kauffman, S.L. & Andersen, D.H. Persistent venous valves, maldevelopment of the right heart, and coronary artery-ventricular communications. *Amer. Heart J.*, 1963, 66, 664-669.
- 21) Kettler, L. Eine seltene, praktisch bedeutsame Vorhofmissbildung. *Z. Kreisl.-Forsch.*, 1934, 26, 649-659.
- 22) Kjellberg, S.R. *Diagnosis of Congenital Heart Disease*, translated by Erica Odelbert, Chicago, The Year Book Medical Publishers, Inc., 1955, p. 25.
- 23) Lauenstein, C. Varietät der Klappen des rechten Atriums. *Virchows Arch. path. Anat.*, 1876, 68, 632-633.
- 24) Le Count, E.R. Network Formations in the right auricle, with demonstration of a specimen. *Trans. Chic. path. Soc.*, 1901-1903, 5, 309-213.
- 25) Leo, H. Über einen Fall von Entwicklungshemmung des Herzens. *Virchows Arch. path. Anat.*, 1886, 103, 503-515.
- 26) Lindes, G. Ein Beitrag zur Entwicklungsgeschichte des Herzens, Inaug. Dis., Doprát. 1865 (cited by Yater in 49).
- 27) Looser, E. Ueber die Netzbildungen im rechten Vorhofe des Herzens. Inaug. Dis. Zürich. 1902 (cited by Yater in 49 and von Moellendorff in 42).
- 28) Lucas, R.V., Jr. & Schmidt, R.E. *Heart Disease in Infants, Children and Adolescents*. Edited by A.J. Moss & F.H. Adams. The Williams & Wilkins Co., Baltimore, 1968, pp. 672-727.
- 29) Maier, R. Zur Casiustik der Herzfehler. *Ber. Verh. nat. Ges.*, 1867, 4, 478 (cited by Yater in 49).
- 30) Moore, N. Variety in the Structure of the Heart, *Trans. path. Soc., Lond.*, 1883, 34, 31-32.
- 31) Ponhold, J. Ein Cor triatriatum biventriculare. *Frankfurt. Z. Path.*, 1937, 51, 237-241.
- 32) Preisz, H. Beiträge zur Lehre von den angeborenen Herzanomalien., *Beitr. path. Anat.*, 1890, 7, 245-296.
- 33) Przewoski, E. Anomaliae chordae cordis humani; valvule venae cavae superioris. *Pam. Towarz. Lek. Warszaw*, 1896, 92, 400 (cited by Yater in 49).
- 34) Röse, C. Beiträge zur vergleichenden Anatomie des Herzens der Wirbelthiere. *Morphol. Jahrb.*, 1890, 16, 27 (cited by Yater in 49).
- 35) Rossall, R.E. & Caldwell, R.A. Obstruction of inferior vena cava by a persistent Eustachian valve in a young adult. *J. clin. Path.*, 1957, 10, 40-45.
- 36) Ruggieri, A. Über eine ausgedehnte Diaphragmabildung im rechten Herzvorhof. *Zbl. allg. Path.*, 1935, 63, 129-133.
- 37) Sabatier. *Mém. Acad. Roy. Sci. Paris*, 198. (1774) (cited by Hickie in 18).
- 38) Scholmerich, P., Stein, E., Klinner, W. & Zenker, R. Transposition der unteren Hohlvene mit Zyanose und Linkshypertrophie. *Verh. dtsh. Ges. Kreisl.-Forsch.*, 1962, 28, 321.
- 39) Stadler, O. Über eine seltene Missbildung des Herzens. *Verh. phys.-med. Ges.*, 1890-1891, 24, 61 (cited by Yater in 49).
- 40) Sternberg, C. Beiträge zur Herzpathologie. *Verh. dtsh. path. Ges.*, 1913, 16, 253-262.

- 41) Thilo, L. Zur Kenntnis der Missbildungen des Herzens. Inaug. Dis. Leipzig. 1909 (cited by Yater in 49).
 - 42) Von Moellendorff, W. Ueber abnorme Erhaltung der Sinuskappen im rechten Vorhof eines menschlichen Herzens. *Anat. Anz.*, 1912, **40**, 406-409.
 - 43) Von Rokitansky. *Die Defecte der Scheidewände des Herzens*. Vienna, 1875 (cited by Yater in 49).
 - 44) Weber, F.P. Interesting cases in which a so-called Chiari's net was found in the right auricle of the heart, with or without the presence of any other congenital cardiac abnormality. *Intern. Clin.*, 1920, **3**, 43-51.
 - 45) Weber, A. Restes de la valvule veineuse gauche dans le coeur humain adulte. *Bibliog. anat.*, 1904, **13**, 11 (cited by Yater in 49).
 - 46) Winslow, J.B. *Mém. Acad. roy. Sci. Paris*, 211. (1717) (cited by Hickie in 18).
 - 47) Wortmann, W. Ueber eine seltene Herzmissbildung. Zugleich ein Beitrag zur Frage der Netzbildungen im rechten Vorhofe. Inaug. Dis. Wurzburg., 1909 (cited by Yater in 49).
 - 48) Wurm, H. Angeborener Herzfehler mit "korrigierter" Transposition der grossen Gefaesse. *Virchows Arch. path. Anat.*, 1927, **263**, 123-141.
 - 49) Yater, W.M. Variations and anomalies of the venous valves of the right atrium of the human heart. *Arch. Path.*, 1929, **7**, 418-441.
-