Constrictive Pericarditis with Atrial Septal Defect in Children

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SUMMARY

Two cases (1-year-old and 13-year-old) of constrictive pericarditis (CP) associated with atrial septal defect (ASD) are presented. The correct diagnosis was made preoperatively in both cases. Both patients underwent surgery (pericardiectomy and ASD closure). One patient died soon after the surgery because of low cardiac output. The other patient was associated with protein-losing enteropathy and it improved dramatically after the operation. Cardiac catheterization is necessary for the diagnosis of this unusual combination. The cause of this unusual combination is obscure.

Additional Indexing Words :

Constrictive pericarditis Atrial septal defect Protein-losing enteropathy

ATRIAL septal defect is one of the most common congenital malformations of the heart. Constrictive pericarditis is less common in childhood especially under the age of 10.¹⁾ The association of CP with ASD is extremely rare and only 12 cases²⁾⁻⁸⁾ have been reported. We recently studied 2 cases of CP associated with ASD in pediatric age group. Most of the patients reported previously are adult. These 2 cases of ours are the youngest patients with this unusual combination.

CASE REPORT

Case 1.

The patient was well until 5 months old when he developed poor feeding, fever, and cyanosis. He was admitted to another hospital, where chest roentgenogram revealed totally opaque left lung field. Thoracentesis revealed hemothorax. A week later he developed chylothorax and was treated with medium chain triglyceride (MCT). He, however, developed congestive heart failure and

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was referred to Nihon University Hospital on Sept. 11, 1975. Physical examination revealed poorly nourished child and facial edema was noted. The pulse was regular, 152/min, respiration, 66/min and blood pressure 110/62 mmHg without paradoxical pulse. There was a grade 3/6 harsh ejection type systolic murmur along left sternal border. A grade 2/6 diastolic rumble was heard at lower left sternal border. Second sound was narrowly split. A liver edge was felt 2 cm below the right costal margin. The electrocardiogram (Fig. 1) revealed normal sinus rhythm, right ventricular hypertrophy, diminished left ventricular forces and diffuse ST-T changes. Chest rentogenogram (Fig. 2) showed moderate cardio-

Fig. 1. Case 1. Electrocardiogram.

There are marked right axis deviation, right ventricular hypertrophy, mild low voltage in limb leads and ST-T changes in all limb and precordial leads.

Fig. 2. Case 1. Chest rentogenogram.

The heart size is moderately increased. There is a sign of increased hilar markings suggestive of left to right shunt.

megaly, pulmonary congestion, and pleural adhesion. Sodium restriction, digitalis, and diuretics were instituted and congestive heart failure was improved. Cardiac catheterization on Sept. 26, 1975 (Table I) showed a left to right shunt at the atrial level, elevated right atrial pressure, mild pulmonary hypertension, and systolic gradient of 20 mmHg across the pulmonary valve. This gradient was thought to be due to a flow related pulmonary stenosis. Fluoroscopy revealed poor cardiac motion and presence of pericardial effusion was suspected. Pericardiocentesis was performed and 45 ml of serosanguineous fluid was removed. A month later the patient was discharged.

On Jan. 23, 1976, he was readmitted because of severe congestive heart failure with left pleural effusion. The patient was treated with digitalis and diuretics and was discharged 2 weeks later. On Aug. 16, 1976 he was admitted again because of poor feeding and dehydration. Electrocardiogram showed atrial fibrillation. He was complicated with disseminated intravascular coagulation and was treated with heparin. Despite vigorous treatment for congestive heart failure, it didn't improve. Echocardiogram on Oct. 29, 1976 revealed no pericardial effusion but poor left ventricular contractility. He was thought to have CP and repeat cardiac catheterization was performed on Nov. 18, 1976. It revealed again a left to right shunt at atrial level. Right atrial mean pressure was markedly elevated and a diastolic right ventricular "dip" and plateau was noted. Pulmonary artery was not entered. On Nov. 30, 1976 the patient was transferred to the operating room. At thoracotomy, CP with thick adhesion over both ventricle was found. Pericardial resection was done, and then a 3x3 cm secundum type ASD was closed. He, however, dropped into low cardiac output state and died soon after the operation. Autopsy was not performed.

Case 2.

A 13-year-old girl was referred to Nihon University Hospital on July 7, 1975

Table I. Cardiac Catheterization Data (Case 1)

	O ₂ Saturations (%)	Pressures (mmHg)
Superior vena cava	72	
Right atrium	81	a = 12 v = 12 m = 9
Right ventricle	89	65/15
Pulm. artery	89	45/15 m = 30
Left atrium	93	a = 15 v = 12 m = 10
Flow (L/min/M ²)		
Pulmonary (Q _p)		9.5
Systemic (Q _s)		5.0
Pulmonary resistance (Unit)		2.1
Q _p /Q _s		1.9

because of edema. A heart murmur was first noted at the age of 7 in another hospital, and clinical diagnosis of ventricular septal defect was made. She had been asymptomatic until 1 week prior to admission when she first noticed edema on legs. Physical examination revealed a well nourished and developed girl. The pulse was regular, 82/min, respiration, 22/min and blood pressure, 118/62 mmHg without paradoxical pulse.

Jugular vein was distended. There was a grade 3/6 harsh ejection type systolic murmur, best heard at upper left sternal border, followed by a fixed wide split second sound. No diastolic rumble was heard. Faint third sound was present at the apex. No thrill was felt. A non-tender liver edge was felt 4 cm below the

Fig. 3. Case 2. Electrocardiogram.

(A) Preoperative: Note T wave inversion in II, III, aVF, and V₁~V₆, and right ventricular hypertrophy.

(B) Eight months postoperative: Note positive T wave in II, III, aVF, and V₅~V₆. Right ventricular hypertrophy still remained.

right costal margin. The presence of ascites was suspected, and abdominal mass was felt at the lower abdomen.

The electrocardiogram (Fig. 3A) revealed normal sinus rhythm, T wave in-

Fig. 4. Case 2. Chest rentgenogram.

The heart is moderately enlarged. The pulmonary vascular marking is increased. The pulmonary artery segment is prominent.

Fig. 5. Case 2. Echocardiogram (preoperative).

Note increased right ventricular dimension and paradoxical interventricular septal motion. There is a sign of minimal pericardial effusion.

RV=right ventricle; IVS=interventricular septum;

TV=tricuspid valve; MV=mitral valve;

LVPW=left ventricular posterior wall; EKG=electrocardiogram.

version in leads II, III, aVf and chest leads. No low voltage was present.

Chest roentgenogram (Fig. 4) showed cardiomegaly and increased pulmonary flow. There was no calcification. Echocardiogram (Fig. 5) revealed increased right ventricular dimension, paradoxical motion of the interventricular septum and minimal pericardial effusion.

Serum protein on admission was 6.0 Gm/100 ml. It, however, decreased to below 5.0 Gm/100 ml a few days after the admission. Liver function tests were normal.

Cardiac catheterization on Aug. 20, 1975 (Table II) showed elevated right atrial pressure, a left to right shunt at the atrial level, mild pulmonary artery hypertension, and systolic gradient of 30 mmHg across the pulmonary valve. A diastolic "dip" and plateau right ventricular pressure curve was noted (Fig. 6A).

The patient was placed on digoxin and diuretics. Plasma and albumin was given intravenously for the treatment of hypoproteinemia every day. The serum protein level, however, did not elevate. ^{131}I -labeled polyvinylpyrrolidone (PVP) was given intravenously to find protein-losing enteropathy. Fecal output of ^{131}I -PVP was 25%, indicating the presence of protein-losing enteropathy.

The abdominal tumor was thought to be the cause of protein-losing enteropathy and laparotomy was performed on Feb. 21, 1976. The tumor was benign ovarian cystic teratoma. In spite of ovarian tumor resection, ascites reaccumulated and serum protein remained low. The patient was thought to have CP and was referred to cardiac surgery. On April 4, 1976 operation was performed. The peri-

Table II. Cardiac Catheterization Data (Case 2)

	Preoperative (8/2/75)		Postoperative (12/3/76)	
	O ₂ Saturations (%)	Pressures (mmHg)	O ₂ Saturations (%)	Pressures (mmHg)
Superior vena cava	75	a=14 v=10 m=12	72	
Right atrium	87	a=14 v=8 m=12	72	a=5 v=6 m=3
Right ventricle	86	70/10	74	40/4
Pulm. artery	89	40/15 m=25	74	32/12 m=19
Left atrium	96	a=8 v=12 m=10	95	a=10 v=12 m=6
Left ventricle	96	125/15	95	126/12
Flow (L/min/m ²)				
Pulmonary (Qp)	10.5		3.6	
Systemic (Qs)	3.5		3.5	
Pulmonary resistance (Unit)	1.4		3.6	
Qp/Qs	3.0		1.0

Fig. 6. Case 2. Right ventricular pressure curve.
(A) Preoperative: Note elevated right ventricular pressure and a diastolic "dip" and plateau curve.
(B) Eight months postoperative: Right ventricular pressure decreased. A "dip" and plateau curve still remained.

Fig. 7. Change of serum total protein.

cardium was found to be markedly thickened (3 mm thick). Pericardiectomy was successfully done, and a 3×4 cm secundum type atrial septal defect was closed with the aid of extracorporeal circulation. Postoperatively the patient was complicated with transient atrial flutter, but signs of congestive heart failure reduced gradually with digitals and diuretic therapy. Intravenous albumin administration was continued after the operation. Three weeks after the operation, serum protein level rose above 7.0 mg/100 ml and intravenous albumin was discontinued (Fig. 7). Serum protein level remained above 7.0 Gm/100 ml and the signs of ascites and congestive heart failure disappeared. She was discharged on Aug. 20, 1976.

Eight months after the operation repeat cardiac catheterization was carried out on December 3, 1976 (Table II). The catheter was entered through the atrial septum to the left atrium. No significant shunt at the atrial level, however, was demonstrated. Right atrial and right ventricular end-diastolic pressure were normal, but a right ventricular diastolic "dip" and plateau curve was still remained (Fig. 6B).

DISCUSSION

Constrictive pericarditis is far less common in children than adult, and is particularly rare in young children. In our pediatric department, 3 out

Table III. Summary

No.	Author	Year	Age	Sex	Type of ASD	Systolic murmur	Diastolic rumble
1	Gignoux*	1910	?	M	2°	?	?
2	Quirno and Battro*	1936	22	M	2°	?	?
3	Semler and coworkers ²⁾	1960	33	M	2°	+	—
4		1960	17	M	2°	+	—
5	Mihailescu*	1962	46	F	2°	?	?
6	Yurchak and coworkers ³⁾	1965	68	M	2°	+	+
7	Yahini and coworkers ⁴⁾	1966	51	M	2°	+	—
8	Just and Mattingly ⁵⁾	1967	27	M	2°	?	?
9	Albers and coworkers ⁶⁾	1969	40	M	2°	+	—
10		1969	14	M	2°	+	—
11	Kumar and coworkers ⁷⁾	1971	65	M	2°	+	+
12	Kotoda and coworkers ⁸⁾	1972	28	M	2°	—	—
13	Harada and coworkers		1	M	2°	+	+
14			13	F	2°	+	+

*=Quoted from Just's paper⁵⁾; 2°=Secundum type atrial septal defect; FS=fixed split; NS=

of 25 cases of pericarditis developed constriction. Two of them are the reported cases in this paper. One is tuberculous CP. The incidence of CP is 12% of all cases of pericarditis. Reviews on pericarditis in children have shown an incidence of constriction ranging from 0.7 to 13%.^{9),10)}

The association of pericardial disease and congenital heart malformation has been rarely reported in the past.^{2),11)} The combination of CP and ASD is quite rare and to the best of our knowledge, only 12 cases have been reported (Table III). Most of them are adults. A 14-year-old patient was reported by Albers et al.⁶⁾ This is the only patient in pediatric age group. Case 1 in this report is the youngest on record.

Constrictive pericarditis is predominantly a disease of male. In reviewing the sex incidence of CP with ASD, only one female case has been reported. Our patient (Case 2) is the second female case on record.

Although CP is rare in association with congenital malformations of the heart, ASD has the highest tendency to associate with pericardial disease. Just and Mattingly⁵⁾ reviewed 67 cases of pericardial disease associated with ASD. They described that 12% of autopsy cases of ASD had some degree of

of Cases Reviewed

Second sound	Para-doxical pulse	Ascites	Hepato-megaly	Edema	Cardio-megaly	Calcification	Friction rub	Operation	Outcome
?	?	?	?	?	?	?	?	?	?
?	?	?	?	?	?	?	?	?	?
NS	?	?	?	?	+	+	?	+	S
NS	?	+	+	?	+	?	?	+	S
?	?	?	?	?	?	?	?	?	?
FS	?	—	+	?	+	+	+	+	S
FS	—	—	+	—	+	—	—	+	D
?	?	?	?	?	?	?	?	+	S
NS	—	?	+	+	+	+	—	+	S
FS	—	+	+	+	+	?	+	+	S
NS	?	+	+	+	+	+	?	—	D
FS	—	+	+	+	+	—	—	+	S
NS	—	+	+	+	+	—	—	+	D
FS	—	+	+	+	+	—	—	+	S

normally split; D=died; S=survived

pericardial disease. To our knowledge, there is no such a tendency in other congenital malformations of the heart. In reviewing previously reported cases and our cases, only secundum type ASD associates with CP, although pericardial effusion is demonstrated in primum type as well as secundum type ASD. Why does pericardial effusion occur in association with ASD? One of the explanations is that elevation of right atrial pressure caused by a left to right shunt at the atrial level compress the coronary sinus, followed by pericardial effusion. Experimental work suggests that compression of the coronary sinus results in accumulation of pericardial effusion.¹²⁾ The true explanation, however, remains obscure.

It is well known that CP accompanies hypoproteinemia.¹³⁾ All the previously reported cases with CP and ASD are not associated with hypoproteinemia. Our patient (Case 2) is the first case complicated with CP, ASD, and hypoproteinemia. Usually CP produces ascites and facial edema and a clinical picture resembles nephrotic syndrome. It is called "pseudonephrosis". Protein-losing enteropathy is the major cause of hypoproteinemia in CP. Hypoproteinemia may also be the result of impaired liver function followed by impaired protein production, loss of protein to the ascites or increased plasma volume by dilution.¹⁵⁾ In our patient, the abnormal permeability of the gut to serum protein was studied by the intravenous administration of ¹³¹I-PVP.¹⁴⁾ The loss of ¹³¹I-PVP to the feces was markedly elevated. The liver function tests were normal. Therefore we concluded that the cause of hypoproteinemia of this patient was loss of protein to the feces. Since this patient had ovarian tumor, it misled the cause of protein-losing enteropathy.

The relationship between protein-losing enteropathy and congestive heart failure has been discussed.¹⁵⁾ Davidson et al studied 3 cases of chronic constrictive pericarditis and ASD with congestive heart failure utilizing intravenous ¹³¹I-PVP and ¹³¹I-albumin. The permeability of the gut to ¹³¹I-PVP was higher than normal. ¹³¹I-albumin turnover studies indicated that hypoalbuminemia was related to subnormal body albumin content and this was the result of an increased rate of albumin catabolism, and not of a failure of albumin synthesis. Why does congestive heart failure predispose gastrointestinal system to loss of plasma protein? A hypothesis is that the elevated venous pressure leads to lymphatic congestion of the intestine as a result of impaired venous return.¹⁶⁾ This fact can give rise to loss of protein through the intestinal wall and result in protein-losing enteropathy. In other cardiac diseases, however, it is extremely rare to cause protein-losing enteropathy even being accompanied by severe congestive heart failure. Constrictive pericarditis might have some specific reason to predispose gastrointestinal system to

loss of plasma protein.

The clinical diagnosis of ASD is relatively easily suspected. Physical examination and echocardiogram play a major role for the diagnosis of ASD.

Echocardiographic features of ASD was well defined.^{17)–20)} Paradoxical interventricular septal motion and increased right ventricular dimension are the echocardiographic findings of right ventricular volume overloading. Pool et al²¹⁾ studied the septal motion of CP and found that the interventricular septum moves paradoxically. They concluded that the septal motion of CP was indistinguishable from paradoxical motion associated with right ventricular volume overloading. Gibson et al²²⁾ also mentioned abnormal septal motion in CP. This abnormal septal motion is also observed in left bundle branch block^{23),24)} and septal damage.^{25),26)} Since left bundle branch block is rare in children, paradoxical septal motion in echocardiogram suggests right ventricular volume overloading and/or CP. In association with CP and ASD, echocardiogram is not diagnostic of either ASD or CP. Since it is very rare to develop congestive heart failure in children with ASD, echocardiographic paradoxical septal motion with the signs of severe congestive heart failure suggests the possibility of CP.

Although the signs of ascites, venous distention, hepatomegaly and peripheral edema were noted, paradoxical pulse was not observed in either patient. Paradoxical pulse was almost invariably present in the majority of cases of CP in Wood's series.²⁷⁾ It was not observed in Albers⁶⁾ and Yahini's cases⁴⁾ as well as our cases. Paradoxical pulse depends on respiratory variations of systemic venous return to the right side of the heart. If right ventricular output was kept constant, paradoxical pulse could not be produced despite an increase in pericardial pressure. In presence of ASD, the blood volume in the right side of the heart remains constant during inspiration and expiration. This is probably the reason why paradoxical pulse is absent in patients with CP and ASD.

Cardiac catheterization is the crucial procedure for the diagnosis of CP. Elevated venous pressure and M shaped (steep X and Y descents) atrial pressure curve is characteristic.⁶⁾ A diastolic "dip" and plateau curve in right ventricle is diagnostic. Our patient's postoperative cardiac catheterization, repeated 8 months after the operation, revealed a right ventricular "dip" and plateau curve, although she was clinically asymptomatic. Myocardial atrophy has been demonstrated in patients with CP.²⁸⁾ Probably this is the factor considered to be responsible for persistent right ventricular "dip" and plateau curve.

Constriction in children is not necessarily a chronic disease as in adult. In our patient (Case 1), constriction developed at most within 6 months after

effusive pericarditis. Rapidly developed CP has been reported by other authors. Strauss et al¹⁰⁾ reported a case in which CP developed only within 8 weeks.

The similarities of CP and cardiomyopathy has been emphasized.^{6),27),29)} In cardiomyopathy, the left atrial and/or pulmonary capillary wedge pressure has been found to be higher than right atrial pressure. In CP, both atrial pressures are elevated to an equal level. In the presence of ASD, both atrial pressures tend to be equalized. This fact makes this differentiation more difficult. Constrictive pericarditis is a correctable disease. Cardiomyopathy, however, is not a correctable disease. Therefore the differential diagnosis of these diseases is of great importance.

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