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Anatomical Consideration in the Surgical Treatment of Type III Ventricular Septal Defect (Atrioventricular Canal Type)

Michio Yokota, Ryusuke Muraoka, Masataka Osaragi,
Minoru Aoshima, Inshin Kyoku, Shinichi Nomoto,
Hiroyuki Nakano, Ken Ueda and Akihiro Saito

Department of Cardiovascular Surgery and Department of Pediatric Cardiology,
Shizuoka Children's Hospital
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Incidence of ventricular septal defect of type Ⅲ by Kirklin's classification (Fig. 1)[11] varies considerably with the series reported[1)2)3)5)8)9)11)14)15)18)19)21)], presumably due to the difference of recognition of the lesion at the time of surgery. The fact is that the described definition of the classifcation lacks a definte anatomical landmark. It has been stated that closure of the defect of this type requires a particular attention in order to avoid the injury to the conduction system. Technically, the insertion of a patch under the bundle of chordae which often crosses the center of the defect sometimes becomes a problem. The purpose of this paper is to report on the surgical and autopsied patients in our clinic who have type Ⅲ defects.

Material and method

The series comprises 14 operated patients and 2 non-surgical autopsied patients with type Ⅲ defect who were encounted at Shizuoka Children's Hospital since its opening of July, 1977. Characteristic feature in this series was that 7 of 14 operated patients and both autopsied patients had Down syndrome. The operated patients are listed in Table 1. The age at the time of surgery ranged from 7 months to 8 years and 3 months with an average of 2 years and 8 months, and body weight ranged from 5.4 kg to 18.0 kg with an average of 10.2 kg .Associated anomalies were Down syndrome in 7 patients, patent ductus arteriosus in 3, and slight mitral regurgitation in 2. On electrocardiogram 7 of 14 patients showed left axis deviation and 6 incomplete right bundle branch block preoperatively. Cardiac catheterization showed moderate and severe pulmonary hypertension (pulmonary arterial pressure to systemic blood pressure in systole higher than 0.6) in 9 patients. In 5 patients localization of the defect was possible and greatly suspected in 3 by angiocardiography on the finding of the low position of the jet through the defect which was interpreted as far

Key words : ventricular septal defect, Type III ventricular septal defect, Atrioventricular canal type VSD, Down syndrome.
索引語：心室中隔欠損症，Ⅲ型心室中隔欠損症，房室口型心室中隔欠損症，ダウン症候群.
Present address : Michio Yokota, Department of Cardiovascular Surgery, Shizuoka Children's Hospital, 860 Urushiyama, Shizuoka, 420, Japan.

Table 1. Summary of operative cases

	cases	associated anomalies	Pp/Ps	magnitude of shunt (of Q_{PA})	mean electric axis (preop)	type diagnosed by Angio.	operative findings	method of closure	outcome	postop. ECG (RBBB)
1	H.K. 2y. 7m.	—	0.33	64%	+50° *	Yes	 2 m 4 m 5 m	direct	alive	(+) compl.
2	E.I. 8y. 3m.	—	0.33	35%	Superior axis by VCG	No	 4×5mm	direct	alive	(−)
3	Y.O. 3y. 11m.	Down PDA	0.96	32%	−30°	Yes	 12×10mm	patch	alive	(+) compl.
4	N.M. 1y. 1m.	Down sl. MR	0.64	43%	+75°	No	 5×4mm	direct	alive	(+) incompl.
5	K.N. 2y. 3m.	Down	0.95	54%	Superior axis * by VCG	Yes	 20×15mm Ⅱ + Ⅲ	patch	alive	(+) incompl.
6	H.I. 2y. 11m.	Down	0.92	L-R(of Q_{PA}) 38% R-L(of Q5) 23%	+80° *	Suspected	 14×13mm Ⅱ + Ⅲ	patch	alive	(+) compl.
7	T.S. 1y. 11m.	Down sl. MR	1.0	67%	+110°	No	 12×12mm Ⅱ + Ⅲ	patch	alive	(+) compl.

8	Y.O. 2y. 9m.	—	0.48	27%	+80°	Yes		9×8m	patch	alive	(+) compl.
9	M.A. 1y. 1m.	Down PDA	1.0	61%	undetermined*	No		10×9mm Ⅱ + Ⅲ	patch	alive	(+) incompl.
10	Y.M. 9m.	—	0.88	63%	−40°	Suspected		11×12mm Ⅱ + Ⅲ	patch	dead (LOS)	—
11	K.Y. 3y. 6m	—	1.0	32%	+100°*	No		14×15mm Ⅲ	patch	alive	(+) incompl.
12	D.T. 7m.	PDA	0.5	40%	−120°	Suspected		10×10mm Ⅱ + Ⅲ	patch	alive	(+) incompl.
13	K.T. 1y. 6m.	—	0.46	70%	−0°	No		φ 7 mm Ⅲ	patch	alive	(+) compl.
14	S.K. 4y. 3m.	Down	1.0	50%	−100°*	Yes		φ 15mm Ⅲ	patch	alive	(+) incompl.

* incomplete RBBB

MR : mitral regurgitatin, Q_{PA} : pulmonary blood feow, Qs : systemic blood flow, RBBB : right bundle branch block, LOS : low cardiac out put syndrome.

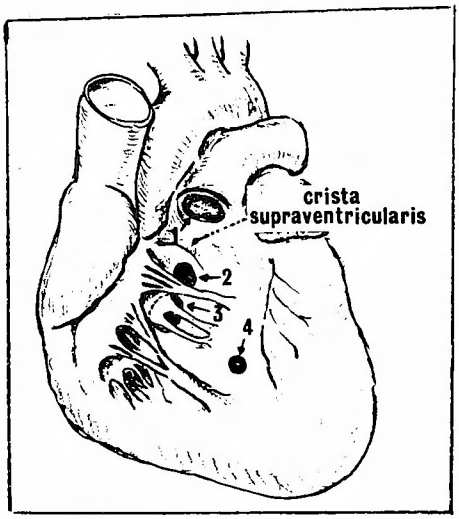


Fig. 1. Classification of ventricular septal defects by Kirklin and associates. Numerals designate each type described in the text. (Redrawn from J.W. Kirklin, and associates, J. Thorac. Surg. 33 : 45, 1957)[11]

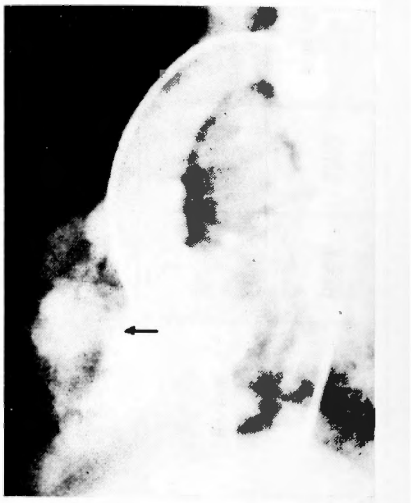


Fig. 2. Left ventriculogram showing low position of the jet through the defect.

below the aortic valve (Figure 2).

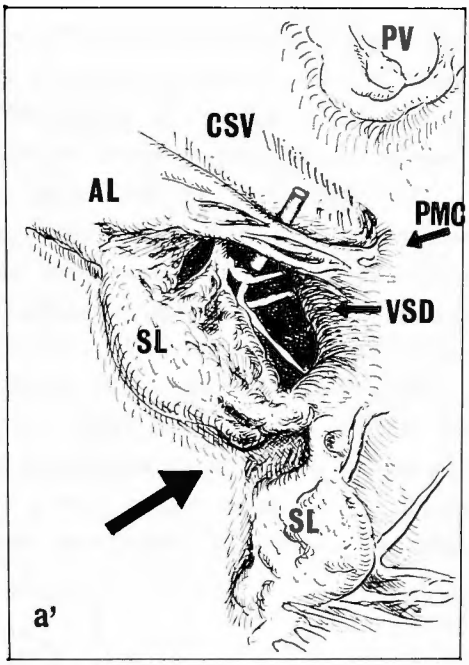
Pathology of autopsied specimens

In two specimens of the non-surgical autopsied patients the ventricular septal defect was the sole cardiac anomaly. On the right ventricular side, the defect was located below the crista supraventricularis and beneath the septal leaflet of the tricuspid valve which composed the posterior margin of the defect. Back side of the septal leaflet of the tricuspid valve attached to the inferior edge of the defect approximately in its midway between the commisure of the anterior and septal leaflets and that of the septal and posterior leaflet. The inferior margin of the defert was situated far caudally to the papillary muscle of the conus which crossed the upper portion of the defect (Fig. 3-a, a′). On the left ventricular side, the defect was in the posterior septum involving the area of the membranous septum extending toward the mitral valve (Fig. 3-b). The defects of these two specimens were interpreted as type Ⅱ + Ⅲ by KIRKLIN's classification.

Operative findings and technique

Operative findings in 14 patients were summarized in Table 1. In cases 1 and 2 the edge of the defect was adhered to the perforation of the septal leaflet of the tricuspid valve, and the defect itself was situated far below the papillary muscle of the conus. The defects in these two patients took the form of the left ventricular to the right atrial communication, however, on angiography there was no shunt between the left ventricle and the right

atrium, presumabley due to the coaptation of the leaflets of the tricuspid valve occurring close to the annulus proximal to the defect. In the other patients the defets were located beneath the septal leaflet of the tricuspid valve extending caudally away from the papillary muscle of the conus. In the cases 5, 6, 7, 9, 10, and 12 the area near the membranous



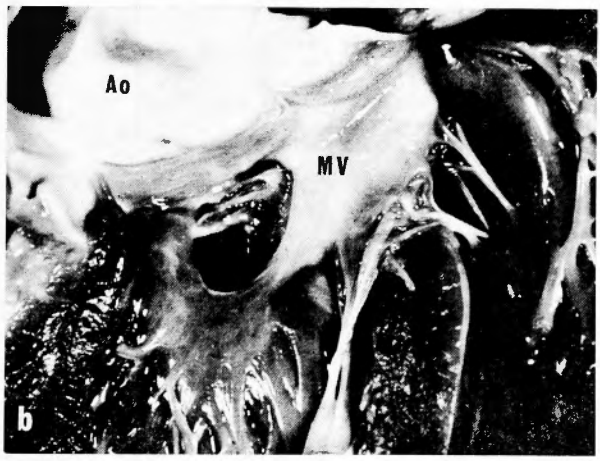


Fig. 3-a, a', b. Autopsied heart specimen with Down syndrome showing type II + III defect, most of which is positioned below the papillary muscle of the conus (small arrow) and attachment of the septal tricuspid leaflet to the inferior edge of the defect simulating a cleft of the septal leaflet (large arrow) in the right ventricular view (a, a'). Aortic valve was bicuspid (b). PV : pulmonary valve, CSV : crista supraventricularis, PMC : papillary muscle of the conus, SL : tricuspid septal leaflet, VSD : ventricular septal defect.

septum was also involved and the defect was regarded as the combination of type Ⅱ and type Ⅲ of KIRKLIN's classification. As with the autopsied cases there was a characteristic feature common to all the large defects of type Ⅲ and type Ⅱ + Ⅲ except for case 14. Back side of the septal leaflet attached to the inferior edge of the defects in its midway between the commisure of the anterior and septal leaflets and that of the septal and posterior leaflets (Fig 4). When the defect was large, only the right half of the inferior edge was covered by the leaflet. The width of the septal leaflet attaching to the muscular septum varied from approximately one mm to three mm. In the cases 5, 6, and 7, one or more of the chordae of the septal leaflet crossed the defect, most of them being caudal (inferior) to the papillary muscle of the conus.

The defect was reached through a median stenotomy. Cardiopulmonary bypass was established in the usual manner with a left ventricular venting. Cardiac arrest was obtained using cardioplegic solution in most cases. In case 1 the ventricular septal defect was approached through a right atriotomy, however, due to the poor operative field, right ventriculotomy was added. In case 14 the defect was closed through the right atrium without difficulty. In cases 1, 2, and 4, the defects were closed by direct suture, and in the other 11 cases, Dacron patch was used. In cases with bridging chordae the sutures first placed on the patch were pulled underneath the chordae and placed to the superior margin of the defect without cutting the chordae. When the attachment of the septal leaflet was narrow, the sutures were placed underneath the attaching septal leaflet (on the

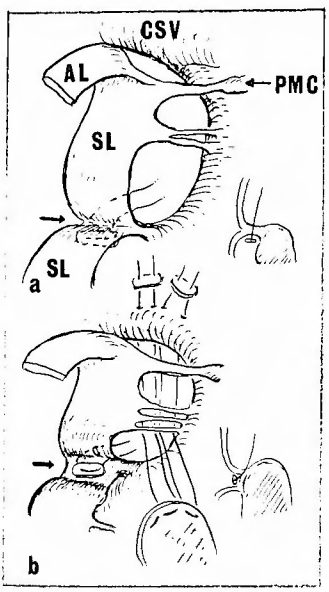


Fig. 4. Placement of the sutures at the posteroinferior corner of the defect (arrows) avoiding the conduction system (VSD type II +III). CSV : crista supraventricularis, AL tricuspid anterior leaflet, SL tricuspid septal leaflet, PMC : papillary muscle of the conus.

right ventricular side) (Fig. 4-a), whereas, when this attachment was wide as in cases 6 and 7, the sutures were placed with the pledgets above the septal leaflet (on the right atrial side) without resulting valvular distortion (Fig. 4-b). Thus the injury of the conduction system was avoided at the postero-inferior corner of the defect using this attaching septal leaflet. The posterior margin of the defect composed by the septal leaflet of the tricuspid valve was closed with the pledgets placed on the right atrial side. In all cases 4-0 Tevdek sutures with pledgets were used.

Results of operation

There was one operative death due to low cardiac output, and no late death. In some patients with Down syndrome the postoperative course was complicated by respiratory disturbance due to the pulmonary vascular disease, and one patient (case 14) had low cardiac output syndrome. There was no complete atrioventricular block in the postoperative electrocardiograms, however, there were incomplete right bundle branch block in 6 patients, and complete right bundle branch block in 6 patients.

Discussion

Various classifications of the ventricular septal defect were advocated so far (Table 2).[2)5)7)9)11)15)16)] While VON ROKITANSKY's classification[16)22)], as modified for the surgeon by WARDEN and associates (1957)[22)], is most complete, the simple anatomic classification of BECU and associates (1956)[2)], modified by KIRKLIN and associates (1957)[11)] is most practical to surgeons and most widely used. Type Ⅲ ("beneath the septal leaflet of the tricuspid valve") is anatomically somewhat vaguely defined in the literatures which caused discrepancies in its incidence reported. (Table 3). In the classification by KIRKLIN and associates[11)] they divided the right ventricular septum into the outflow and inflow tracts, and defined the outflow tract as the portion of the ventricular septum that lies between

Table 2. So-called type Ⅲ defect by the classification of Kirklin and associates appearing in various classifcations.

Rokitansky et al.[16)22)] (1874)	Type B. defect in the posterior septum.
Becu et al.[2)] (1956)	defects involving regions not related to the ventricular outflow tracts, defects related to the atrioventricular valves.
Kirklin et al.[11)] (1957)	defect related to ventricular inflow tracts, beneath septal leaflet of tricuspid valve.
Inomata[7)] (1959)	Type Ⅳ, defect in the posterior ventricular septum.
Neufeld et al.[15)] (1961)	defects of A-V commune type.
Keith[9)] (1965)	A-V canal region VSD, junctional atrioventricular septal defect.
Goor et al.[5)] (1970)	smooth VSD, Type I* and Type II**

* defect of the posterior smooth septum with coincident involvement of septum membranaceum.
** complete absence of the posterior smooth septum.

Table 3. Incidence of the isolated ventricular septal defect by Kirklin's classification

	type Ⅰ	type Ⅱ	type Ⅲ	type Ⅳ	total
Becu et al.[2] (1956)	4	36	6 (12%)	5	51
Warden et al.[22] (1957)	5	71	5 (7%)	3	84
Kirklin et al.[11] (1957)	2	25	8 (21%)	3	38
Neufeld et al.[15] (1961)	···	···	15 (5%)	···	ca. 360
Keith et al.[9] (1967)	8%	70%	8%	12%	
Goor et al.[5] (1970)	13*	63	1 (0.9%)	33%	110
Tatsuno et al.[19] (1970)	32	59	···	2	93
Shozu et al.[18]** (1970)	6	18	5 (17%)	0	29
Kawashima et al.[8] (1971)	40	134	···	···	174
Cooley et al.[3] (1971)	37	573	75 (11%)	17	702
Anzai et al.[1] (1975)	32	64	25 (21%)	0	121
McNicholas et al.[14] (1978)	1	69	4 (5%)	9	83

* including complete defect of the conal septum
** $P_P/P_S > 0.8$

the pulmonary valve above and the nearest part of the tricuspid valve below (or papillary muscle of the conus). They subdivided the defects in the outflow tract into two groups ; one above the crista supraventricularis (type Ⅰ) and one below (type Ⅱ), and also subdivided the defect in the inflow tract into two groups ; one beneath the septal leaflet of the tricuspid valve (type Ⅲ) and one in the muscular portion of the septum (type Ⅳ) (Fig 1). COOLEY and associates[3] actually first named them type Ⅰ, Ⅱ, Ⅲ and Ⅳ accordingly. In the classification of KIRKLIN and associates five major anatomic landmarks of the interior of the right ventricle were paid attention; (1) the tricuspid valve ring and leaflets, (2) the papillary muscle of the conus (of Lancisi), (3) the crista supraventricularis, (4) the annulus fibrosus of the pulmonary valve, and (5) the apex of the ventricular septum. They defined type Ⅱ defect as the one below (inferior to) the crista supraventricularis in the outflow tract. The crista supraventricularis, as seen surgically, is the portion between the papillary muscle of the conus below and posteriorly, and the pulmonary valve above and anteriorly. Therfore, type Ⅱ defect can be defined as one above the papillary muscle of the conus. Type Ⅲ defect which is in the inflow tract can be defined, therefore, as the one below (inferior to) the papillary muscle of the conus and most portion of the defect is covered by the septal leaflet of the tricuspid valve. While they lie in the upper or basal portion of the septum, they are not closely related to the aortic valve when viewed from the left ventricular aspect, and they lie near the posterior leaflet of the mitral valve. Type Ⅲ defect is also called the defect in the posterior septum (ROKITANSKY, 1874)[16][22]. Posterior septum is an embryological terminology and not a purely anatomical one (LEV, 1959)[13-a]. and strictly speaking, all of the septum posterior to the right bundle branch is the posterior septum from the right ventricular aspect (WIDRAN, 1951)[23], and it includes all of the septum posterior to the septum to the septal band of the crista supraventricularis. The

right bundle branch travels at the level of the papillary muscle of the conus, which would reasonablly become an anatomical landmark to delineate the posterior septum.[23] Embryologically, it was demonstrated that the atrioventricular cushions at the base join the muscular part of the ventricular septum to obliterate the interventricular communication in this region (KRAMER, et al. 1941)[12], and final closure of the embryonic interventricular foramen results from fusion of elements of the endocardial cushions of the atrioventricular canal, the conus septum, and the endocardium of the muscular septum (WAKAI and EDWARDS[21] KRAMER et al. 1942[13]). While type Ⅱ defect results from faulty growth of conus ridge, faulty fusion of the endocardial cushions with the conus septum, and the endocardium of the muscular septum causes type Ⅲ defect. KEITH[9] stated that BECU and associates' terminology of inflow and outflow tracts of the right ventricle is open to question since their inflow tract includes defects of the retrocristal junction which is in reality inflow and outflow since it occurs at the junction of both, and also stated that in any classification of ventricular septal defect one must take into acccount the great variety of anomalies that are associated with it. KEITH[9] classified them as "junctional atrioventricular septal defect", which is, however, not practical to surgeons due to its lack of description of anatomical landmarks of the right ventricular septum. KIELY and associates[10] emphasized the possibility that isolated ventricular septal defects of the common atrioventricular canal type may occur with or without involvement of the atrioventricular valves, and in 1961 NEUFELD and associates[15] described 15 such cases anatomically proved. From their description the defects seem to fall into the category of type Ⅲ defects of KIRKLIN's classification when viewed from the right ventricular aspect, namely, the defect extended anteriorly to the crista supraventricularis. A part, at least, of the membranous septum was identifiable and formed the anterosuperior boundary of the defect. The defect did not extend anteriorly beyond the papillary muscle of the conus. The posterior edge of the defect extended more dorsally, and its entire superior edge lay immediately beneath the tricuspid ring. From the left ventricular view, the defect was positioned beneath the membranous portion of the ventricular septum. The upper edge of the defect was separated from the aortic leaflets by the membranous portion of the ventricular septum or by the musculature that corresponds to the crista supraventricularis. In addition deformity or cleft of the atrioventricular valves were notted in 9 of 15 cases. In 1965 EDWARDS and associates[4] divided the aforementioned defect of the persisten A-V canal type or A-V commune type into two groups; posterior and anterior defects. The posterior defect includes the membranous portion of the ventricular septum and extends posteriorly to lie under all of the septal leaflet of the tricuspid valve. The upper edge of the defects is formed by the tricuspid annulus. The anterior variety (Fig. 5) was stated to have some similarity to the common variety of ventricular septal defect (type Ⅱ) which lies posteroinferior to the crista supraventricularis. The exceptional feature of the anterior variety is that it extends downwards against the septal limb of the crista supraventricularis. From the left ventricular aspects such defects extends obliquely downward across the septal wall of the left ventricular outflow tract. This is in contrast to

Fig. 5. Three examples of ventricular septal defect of A-V commune type (anterior variety) lying principally below the papillary muscle of the conus described by Edwards and associates. (Reproduced with permission from Congenital Heart Disease, pages 92 and 93, W. B. Saunders Company, Philadelphia and London)[4].

the usual variety of ventricular septal defect, which in this area is directed horizontally, as it involves the left ventricular outflow tract. They used the papillary muscle of the conus as a definite anatomical landmark to define the anterior variety, namely, the defect of the anterior variety lay principally below or posteroinferiorly to the papillary muscle of the conus.

While the papillary muscle of the conus can be regared as the landmark for type Ⅲ defect, in the series of KIRKLIN and associates[12] nineteen of the twenty five type Ⅱ defects seemed to be completely above the chordae tendineae to the papillary mucle of the conus while six were both above and below these chordae. COOLEY and associates[3] stated that in type Ⅱ defects, the lower portion of the defect were sometimes crossed by the chordae

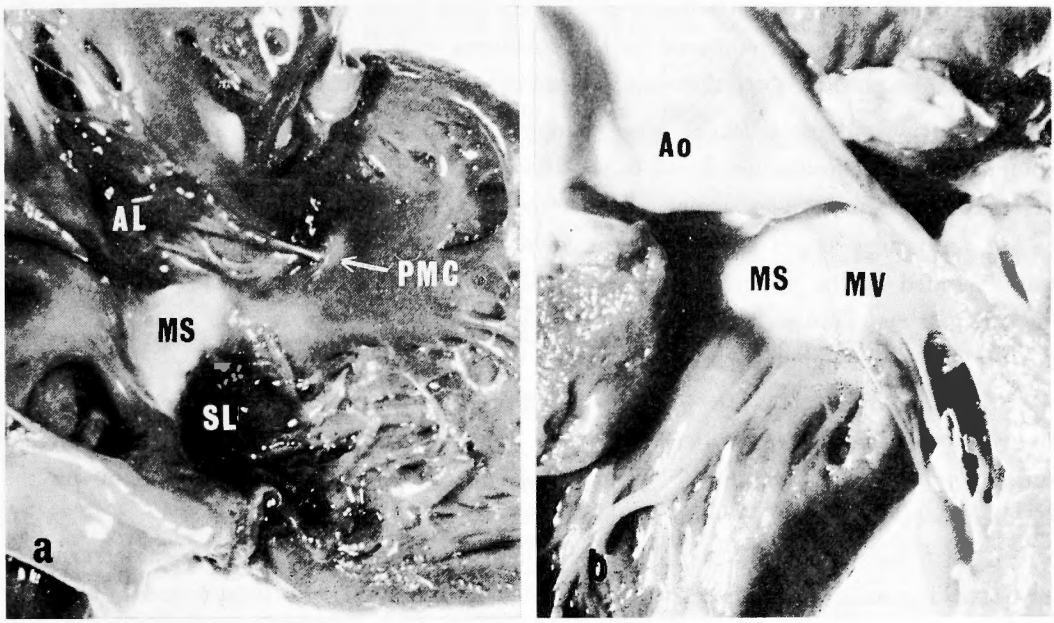


Fig. 6. Autopsied heart specimen without ventricular septal defect in a case with Down syndrome, showing an enlarged membranous septum. (a) Right ventricular view showing also defective superior half of the tricuspid septal leaflet and attachment of the inferior half of the septal leaflet to the inferior edge of the membranous septum. (b) Left ventricular view showing the enlarged membranous septum extending to the posterior mitral leaflet. PMC : papillary muscle of the conus, MS : membranous septum, SL : tricuspid septal leaflet, PV : pulmonary valve, AO : aorta, MV : mitral valve, AL : tricuspid anterior leaflet.

tendineae to the papillary muscle of the conus. In our experience, however, in almost all of the type Ⅱ defect, the chordae tendineae to the papillary muscle of the conus were at the lower edge of the defect, and in all large type Ⅲ defects in this series these chordae were attached to the anterosuperior corner of the defects. Namely, anatomical characteristic features of the large defects in the present series were ; (1) the chordae tandineae to the papillary muscle of the conus were attached to the anterosuperior corner of the defect, (2) in some cases the additional chordae of the septal leaflet crossed the middle of the defect, (3) most portion of the defect were covered by the septal leaflet of the tricuspid valve, and (4) in all except for one case a part of the tricuspid septal leaflet was attached to the right half of the inferior margin of the defect. The latter attachment of the septal leaflet can be seen as a cleft of the septal leaflet when seen from the right atrium (Fig. 3 a,a′). When the portion near the membranous septum above the chordae tendineae to the papillary muscle of the conus is also defective the defect was classified as type Ⅱ + Ⅲ.

Association of Down syndrome with type Ⅲ defects should be emphasized. Seven of 14 in the present series were associated with Down syndrome, and all the ventricular septal defects associated with Down syndrome were type Ⅲ defects in our experience. ROSENQUIST and associates (1974)[17] showed a significant enlargement of the membranous septum in

heart specimens without ventricular septal defect with Down syndrome as compared to the normal heart. We also encountered such a specimen with Down syndrome. From the right ventricular aspect (Fig. 6-a) the membranous portion was enlarged downward. The inferior half of the tricuspid septal leaflet was attached to the inferior margin of the membranous septum, and the chordae tendineae to the papillary muscle of the conus attached to its anterosuperior corner. The superior half of the tricuspid septal leaflet was defective. From the left ventricular aspect (Fig. 6-b) the enlarged membranous septum extended obliquely to be bounded by the posterior mitral leaflet. This raises a question whether the defect of this enlarged membranous portion in the defect with Down syndrome be classified as type Ⅱ or type Ⅲ, although the portion is exactly that of type Ⅱ + Ⅲ defect.

The recongnition of type Ⅲ defect during surgery has the following significance ; (1) closure of the defect is often associated with injury to the conduction system, (2) higher mortality rate with its closure[6)15)], and (3) patch closure is often made difficult by the chordae of the septal leaflet crossing the defect.

NEUFELD and associats[15)] studied the conduction system of the defect of the atrioventricular canal type and noted the course of HIS bundle was similar to those found in the fully developed stage of the malformation as reported by LEV[13-b)], although the position of A-V node was normal in the former and displaced posteriorly in the latter. HIS bundle penetrated the central fibrous body just above the posterosuperior angle of the defect and turned downward posteriorly behind the defect and followed the posterior edge of the defect in the form of an arc. Then the right bundle branch traverses the inferior rim of the defect as in type Ⅱ defect, and further upwards along the anterior rim to the papillary muscle of the conus. The second part of the right bundle branch of HIS courses from the papillary muscle of the conus to the posteroinferior edge of the moderator band (LEV, 1954)[13-c)]. Such long conduction pathway may be the cause of the alteration in excitation described by NEUFELD and associates[15)], TOSCANO BARBARA and DUSHANE[20)], and others. Incidence of the complete right bundle branch block was considerably high with type Ⅲ defect closure, presumably due to the stitches on the anterior margin of the defect. All the defects here were closed through the right ventriculotomy except for the last case (case 14) in which the right ventriculotomy was avoided with the anticipation that the right ventricular pressure would not be sufficiently reduced in the older children with Down syndrome causing postoperative low output syndrome with ventriculotomy. The defect of this type can be closed easily if the chordae tendineae crossing the defect are properly managed.

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和文抄録

Ⅲ型心室中隔欠損症 (Kirklin 分類) の外科的治療

静岡県立こども病院心臓血管外科
横田 通夫，村岡 隆介，大仏 正隆
青嶋 実，曲 人伸，野本 慎一
同 循環器科
中野 博行，上田 憲，斉藤 彰博

共通房室口型ともいわれるKirklin分類Ⅲ型心室中隔欠損症の頻度は報告によって非常に異なるが，解剖学的定義に多少あいまいな点があることから，手術時，ならびに剖検時におけるこの型の識別に差異が生じたものと考えられる．

静岡県立こども病院において，1977年7月の開院以来，Ⅲ型心室中隔欠損症14例の手術および2例の非手術剖検例を経験した．手術例14例中7例および2例の剖検例にダウン症候群を合併していた．この16例中小欠損例2例を除く他の例にみられた解剖学的特徴は，Ⅲ型は，(1) 円錐部乳頭筋 (Lancisi 乳頭筋) が欠損孔の前上縁に附着し，その中隔尖腱索は欠損孔の上縁に接し，時にそれ以外の三尖弁中隔尖の腱索が数本欠損孔を横切っていたこと，また，欠損孔の後縁の殆どの部分が三尖弁中隔尖の弁輪部で形成され，1例を除く全例に欠損孔の下縁の右房側 (後側) 半分に中隔尖の裏面が附着していたことである．この中隔尖の附着は，右房側よりみると中隔尖の裂隙の如く認められた．

我々は，円錐部乳頭筋への腱索が欠損孔の下縁ないし下縁近く走るものをⅡ型，上縁に接しているものをⅢ型，Ⅲ型にこの腱索より上部の膜性部中隔附近の中隔の欠損を合併しているものをⅡ＋Ⅲ型と判定している．

術前心電図上14例中7例に左軸偏位を，6例に不完全右脚ブロックを認めた．

Ⅲ型心室中隔欠損症は，手術死亡率が高く，術中刺激伝導路障害を発生しやすいといわれるが，上記の欠損孔後下縁に附着する三尖弁中隔尖に糸を通すことによって完全房室ブロックは容易に防止しうる．14例中1例を低心拍出量症候群で失なった．6例に不完全右脚ブロック，6例に完全右脚ブロックをみたが完全房室ブロック例はない．欠損孔の中央部を横切る腱索の処理を適切に行なえば，この型の欠損孔は右房から容易に閉鎖しうると考える．

尚，ダウン症候群の膜性中隔が異常に拡大していて同症候群におけるⅢ型中隔欠損の発生機序を示唆する剖検例を示した．