Anatomical-embryological correlates in atrioventricular septal defect

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SUMMARY Recent embryological studies have supported the consideration that the ventricular septum is multifocal in origin. These data have also provided excellent correlation of the morphology of malformed hearts with their embryology. In particular, atrioventricular septal defect correlates accurately with these observations on ventricular septation. Many of the names given to atrioventricular septal defect (for example ostium primum, persistent atrioventricular canal, endocardial cushion defect) indicate attempts at correlating the anatomy with embryology. None of these has been very convincing.

In the light of this uncertainty, this review considers briefly the anatomy of the malformation and its ontogeny, and presents a hypothesis of the development of atrioventricular septal defect.

Although there is almost always a communication above the atrioventricular valves, the malformation lies in the ventricular, not the atrial septum. Hearts with inlet septal defect without interatrial communication represent one end of the spectrum of anomalies, and those with common atrioventricular orifice, in which Fallot's tetralogy or single outlet heart may be associated, mark the other end. The outflow tract malformations are not randomly associated, but are points in a huge range of cardiac malformations.

Atrioventricular septal defect is a cardiac malformation characterised externally by an abnormally short posterior (diaphragmatic) ventricular surface.¹ Internally there is a gap between the concave inferior rim of the atrial septum (which is usually well developed) and the atrioventricular valves. There may be a common atrioventricular valve, usually with five or six leaflets of which the two major ones, anterior and posterior, bridge the ventricular septum.² Often there are two valves, each with three leaflets³ (Fig. 1a, b). The left component has an abnormally orientated anterior leaflet which is separated medially from the septal or posterior leaflet by an unsupported division which is usually called a cleft. Between the anterior and septal leaflets, at the obtuse margin, is the lateral leaflet (Fig. 1b). When there is a bridging leaflet between either the anterior and lateral, or posterior and lateral leaflets, it produces an accessory orifice, and is common in atrioventricular septal defect.^{1,3} The

leaflets themselves are often poorly developed and may have a verrucous appearance.

The short external diaphragmatic surface of the ventricular mass is reflected internally by a pronounced disproportion between the inlet and outlet lengths of the left ventricle. In normal hearts these two measurements are essentially the same.¹ In atrioventricular septal defect the outflow tract of the left ventricle is long and narrow compared with a normal heart (Fig. 2a, b) and the left anterior leaflet inserts immediately behind and beneath the aortic valve so that the latter is displaced. (Whether the valve is really displaced has been debated at length⁴⁻⁶ but the appearance is that of displacement.) It has lost its wedged position between the atrioventricular valves and the ventricular septum.^{7,8} Sometimes aortic outflow tract obstruction occurs, and muscular obstruction may be further aggravated by hypoplasia of the aortic valve.⁹

Pulmonary outflow tract obstruction is especially associated with a common valve with bridging leaflets² and may be the result of the infundibular septal derangement characteristic of Fallot's tetral-

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Fig. 1 (a) The atrioventricular valves in atrioventricular septal defect (AVSD) seen from the right atrium (RA). The atrial septum (AS) is intact and its concave rim forms the "roof" of the AVSD. The valve leaflets are attached at the same level to the summit of the muscular septum, but the anterior (AL) and posterior (PL), or septal (SL) bridging leaflets are separated by the "cleft" (arrowed). LLL, lateral left leaflet; ARL, anterior right leaflet; CS, coronary sinus; FO, fossa ovalis; LFO, limbus fossa ovalis. (b) Same heart, left atrial (LA) view. The left valve does not resemble a normal mitral valve. It has three leaflets, anterior, septal or posterior, and lateral. The abnormally orientated anterior leaflet is separated by the cleft (arrowed) from the septal or posterior bridging leaflet. LAu, left auricle. Other abbreviations as in Fig. 1(a).

Fig. 2 (a) The left ventricular outflow tract in the normal heart. The inlet septum (IS) and its atrioventricular component, indicated by the dotted line, give "depth" to the outflow tract. The atrioventricular septum extends from the membranous part of the septum (MS) to the crux cordis permitting a posterior recess (arrowed). The anterior mitral leaflet (AML) is normally orientated, and extends from the base of the left aortic leaflet (L) to the non-coronary leaflet (NC). Thus the aortic valve is "wedged" by the inlet septum and the mitral valve, so that it lies beside rather than anterior to the mitral valve. Ao, aorta; LVOT, left ventricular outflow tract; R, right aortic leaflet. (b) The left ventricular outflow tract in AVSD. The narrowness is partly the result of the deficiency of the inlet septum and absence of its atrioventricular component (note the septal attachment of the anterior leaflet (AL) of the left valve) and partly of attenuation of the outlet septum (OS). The loss of the normal "wedge" position of the aortic valve is shown by the position of the anterior leaflet which extends from the left aortic leaflet to the commissure between non-coronary and right (R) leaflets. The membranous part of the septum is small, but intact. APM, anterior papillary muscle group. Other abbreviations as in Fig. 1.

ogy. Double outlet right ventricle, ventriculoarterial discordance, and single outlet heart are also sometimes represented in atrioventricular septal defect.¹⁰⁻¹²

The atrial septum is usually well developed in atrioventricular septal defect; the anticipated normal anatomical elements are represented and atrial septation is often complete, especially in those with "partial" or "transitional" defects (Fig. 1a and b). Patency of the foramen ovale, however, occurs with some frequency, as in the normal heart, and secundum atrial septal defects are common. In view of the frequent finding of a normal atrial septum it is, perhaps, surprising that so many investigators have concluded that the development of this structure is at fault in atrioventricular septal defect. The defect itself is the gap between the inferior rim of the atrial septum and the atrioventricular valves. The vertical dimension of this defect is approximately the same as the disproportion between the inlet and outlet length of the ventricular septum measured in the left ventricle (Table 1). These data strongly suggest that the anomaly concerns the inlet ventricular septum rather than the atrial septum.

The foregoing paragraphs are not intended to be an exhaustive description of atrioventricular septal defect, but to indicate the spectrum which can accurately be correlated with the ontogeny of the ventricles and the ventricular septum.

Historical background

Atrioventricular septal defect was first described by Peacock¹³ who considered imperfect ventricular septation to be involved in the anomaly. In view of recent embryological findings, Peacock's view is of some interest, as many of those who followed him considered only defective or arrested septation to be responsible. The history of the terminology and the gross classification are summarised in Table 2.

Table 2 *Nomenclature and morphology in atrioventricular septal defect*

Peacock ¹³ 1846	Imperfection of atrial and ventricular septa
Rokitansky ¹⁴ 1875	Atrioventricular canal defect Partial - VSD Complete + VSD
Watkins and Gross ¹⁶ 1955	Endocardial cushion defect Ostium primum ASD
Wakai and Edwards ¹⁷ 1956	Persistent common AV canal Partial separate atrioventricular valves Transitional almost separate atrioventricular valves ± small VSD Complete common atrioventricular valve
Bedford <i>et al.</i> ¹⁸ 1957	Atrioventricular defect
Brandt <i>et al.</i> ¹⁹ 1972	Endocardial cushion defect Partial - VSD Complete + VSD
Piccoli <i>et al.</i> ^{5,6} 1979	Partial separate atrioventricular valves Complete common atrioventricular valve

ASD, atrial septal defect; VSD, ventricular septal defect.

With respect to "partial" and "complete" defects, most authors have adhered to Rokitansky's¹⁴ division between those with interatrial communication only (partial) and those who had an interventricular defect as well.¹⁵⁻¹⁹ More recently, however, the two terms have been used to distinguish between separated atrioventricular (partial), and common orifice (complete),⁵⁻⁶ but this distinction has not gained general currency, especially in surgical practice.^{2,3} Wakai and Edwards²⁰ described a third type of atrioventricular septal defect which they termed "transitional" or "intermediate". These hearts had two "cleft" atrioventricular valves and a "narrow bridge of valvular tissue in the midline (which) joins the anterior half of each of the two cleft atrioventricular valvular leaflets with their respective posterior halves just above the ventricular septum". In four of the six hearts which they described there was a small interventricular communication as well. Thus the "transi-

Table 1 *Measurements of inlet and outlet septa in left ventricle, and of vertical dimension of defect in 10 hearts with atrioventricular septal defect, situs solitus, and concordant ventriculoarterial connections*

No.	AV valves	Septal length (mm)		Inlet outlet difference (mm)	Vertical dimension of defect (mm)
		Inlet	Outlet		
1	Common	28	48	20	20
2	Separate	30	38	8	10
3	Common	29	41	12	16
4	Almost separate	36	55	19	21
5	Common	36	47	11	12
6	Separate	35	62	27	28
7	Separate	40	54	14	16
8	Common	25	40	15	16
9	Separate	40	65	25	25
10	Common	31	51	20	22

Note: The discrepancy is much the same as the vertical dimension of the defect, and the figures accord with those given by Goor *et al.*,¹ that is that there is about 30% difference between the two. Thus the inlet septum represents about one third of the septal mass.

tional" group are like those with "partial" atrioventricular septal defect because they have two separated valves, but they also resemble "complete" atrioventricular septal defect because they have an interventricular communication.

With respect to the interventricular communication, the authors considered it to represent a deficiency of the membranous part of the septum.^{17 20} This structure, however, is usually present and intact in atrioventricular septal defect though it is abnormally located^{5 6} (Fig. 2b).

(1) VENTRICULAR SEPTUM

In order to correlate the features described, it is necessary to describe the inlet ventricular septum and its atrioventricular component and to review recent embryological studies of ventricular septation.

The inlet ventricular septum is that part of the septum which separates the inlet portions of the left and right ventricles. It is triangular in shape (Fig. 3) and is identified in both ventricles as the smooth septum posterior and superior to the papillary muscles. On the right ventricular surface its boundaries are the tricuspid annulus and the chordal insertion of the septal leaflet of the tricuspid valve. In the left ventricle the boundaries are less well defined as the inlet septum coalesces with the trabecular septum, which extends basally to the membranous part of the ventricular septum.

The atrioventricular septum: the leaflets of both atrioventricular valves are attached to the inlet ventricular septum; because the tricuspid leaflets are attached slightly more towards the apex than those of the mitral valve, a part of the inlet musculature interposes between the right atrium and the left ventricle.

Fig. 3 (a) Diagrammatic outline (from a left ventricular angiocardio-gram) to show the position of the inlet septum. It is wedge-shaped and occupies the area between the mitral valve and the outlet septum (broken lines). (b) The same view in AVSD. The left valve (solid line), notched to indicate gap between the leaflets (the cleft), has "descended" into the left ventricle. The broken lines represent the deficient inlet septum.

This is the muscular atrioventricular septum. It lies below and posterior to the membranous part of the septum, and is distinct from it (Fig. 2a). It extends from its points of coalescence with the trabecular and membranous septa to the crux cordis, producing a well defined posterior recess behind the anterior mitral leaflet. This recess not only gives an anteroposterior dimension to the left ventricular outflow tract but also contributes to the normal wedged position of the aortic valve between the two ventricular inlets and the atrioventricular valves.^{7 8}

Normal cardiac septation: the ventricular septum, as its adult morphology suggests, is multifocal in origin.²¹⁻²⁷ Before septation has begun the inlet and outlet parts of the ventricular loop are distinguishable from one another, and both contribute to the two adult ventricles.²⁴ As the loop elaborates an inlet-outlet foramen becomes recognisable between the two limbs of the loop. This foramen is orientated in such a manner that subsequent septation will place its basal portion in the left ventricle and the apical part in the right ventricle. According to Wenink²⁷ some of the anterior leaflet of the mitral valve elaborates from endocardium from the apical part. The inlet part of the septum develops from loose trabeculations in the posterior wall of the embryonic ventricular mass, while the outlet septum develops from cushions in the outlet part and subsequent infolding of the walls of the outlet.²⁸

By 33 to 34 days (7.5 mm crown-rump) trabeculations appear in the posterior ventricular wall. By 43 days (15 mm crown-rump) these have coalesced to divide the inlet portions of the ventricle. This new inlet septum is obvious at 25 mm (53 days), and by this time the process of undermining of the myocardium which will form the atrioventricular valves and their papillary muscles is under way.²⁶ By 28 mm this undermining is well advanced and the trabecular septomarginalis is emancipated from the septal myocardium. In the adult heart no tensor apparatus is found anterior to the trabecular septomarginalis.

The endocardial cushions, for so long considered to be precursors of the atrioventricular valves and to play a major part in both atrial and ventricular septation,^{4 8} are prominent embryonic cell masses which are first distinguishable at 29 days (3.6 mm).²⁶ They function as valves in the embryonic heart^{29 30} and have regressed by the time (53 days) the atrioventricular valves have elaborated from the ventricular myocardium. This, according to Van Gils,³⁰ is their primary function and they have no role in the septation of the inlets or in forming the adult atrioventricular valves.

(2) ATRIAL SEPTUM

The sequence of events which septate the atria begins fractionally later than that of the ventricles. The sep-

Table 3 *Timing of events in normal cardiac septation*

Gestational age (d)	Crown-rump length (mm)	Event
29–30	3–6	Atrioventricular canal, endocardial cushions, inlet outlet foramen
32	5	Septum primum, ostium primum, endocardial cushions
33–34	7–5	Ventriculobulbar septum, posterior trabeculations visible, right part of atrioventricular canal in direct contact with outlet of future right ventricle
36	10	Ostium primum almost obliterated: septum primum degenerates, ostium secundum formed
43	15	Posterior trabeculations form inlet septum, some begin coalescence to form atrioventricular valves
46	18	Septum secundum meets septum primum anteriorly; septum primum forms foramen ovale
53	25	Endocardial cushions rapidly regressing, myocardial undermining emancipates trabecular septomarginalis and atrioventricular valves
240	Birth	Foramen ovale (septum primum) usually closes

tum primum and endocardial cushions are evident at 32 days (5 mm) and the ostium primum is almost obliterated by 36 days (10 mm).³¹ The septum primum then degenerates to permit a second pathway, the ostium secundum. At about 46 days (18 mm) the septum secundum meets the septum primum anteriorly to form a new opening—the foramen ovale. As is universally recognised, atrial septation is not completed until a short time after birth. Cardiac septation is summarised in Table 3.

Hypothetical development of atrioventricular septal defect

From the foregoing review of normal septation it is possible to hypothesise a developmental background for atrioventricular septal defect.

Although the atrioventricular valve deformities are of supreme surgical importance, in anatomical terms they are secondary to and consequent upon the malformation of the ventricular septum.

DEFECT OF INLET SEPTUM

Abnormal development of the posterior ventricular wall produces a deficiency of the posterior, inlet septum. Because the atrioventricular valves also develop from the posterior myocardium, they are automatically deformed when the posterior septum is deficient. A small degree of posterior maldevelopment might result in a modest defect below and behind the valves, usually called a “canal type defect”.³² Most “canal type defects” are in fact excavated malalignment/perimembranous defects,⁵ but where there is a genuine inlet/outlet discrepancy of the left ventricle,

they are true atrioventricular defects.⁷ A bigger posterior septal deficit would carry the valves down into the ventricles, so the gap (defect) is between them and the floor of the atrial septum (so-called ostium primum atrial septal defect). A huge deficit, readily measured anatomically (Table 1), permits the big interventricular communication extending as far forwards as the anterior septum. The septal deficiency is large enough to leave the valves behind, so that the major leaflets of the common valve float, bridging the septal deficit. Because the anterior mitral leaflet elaborates from the outlet part of the loop, it remains high in the left ventricle, sometimes attached to the posterior rim of the outlet septum and the aortic valve (Fig. 4a, b).

ADDITIONAL ANOMALIES

Because the inlet/outlet foramen develops before the inlet septum, coincidental maldevelopment of this region would just antedate inlet septum deficiency. As the foramen is orientated before its septation, myocardial cells are carried to the basal part of the left ventricle. In normal development these contribute to the anterior mitral leaflet. The same probably occurs in atrioventricular septal defect, permitting attachment of the left valve beneath the aortic valve when the defect is small. If the deficiency of the inlet septum is large, however, then the aortic valve may lose its anchorage in the left ventricle and become a right ventricular structure. Maldevelopment of the outlet septum such as anterior deviation of the outlet (bulbar) cushions would produce infundibular obstruction such as that in Fallot's tetralogy³³ and this may be exacerbated by muscle bands and abnormal placement and development of the trabecula septomarginalis.³⁴

The final morphology of atrioventricular septal defect is determined by two factors—the extent of posterior septal deficiency governs the size of the defect (and probably the valve morphology too) while the degree of maldevelopment of the ventriculo-infundibular cushions dictates the morphology of the outflow tracts.

Correlates

(1) VENTRICULAR SEPTUM IN ATRIOVENTRICULAR SEPTAL DEFECT

The anatomical hallmarks of atrioventricular septal defect are deficiency of the triangular inlet septum and absence of its atrioventricular component. Deficiency of the inlet septum is shown by the short diaphragmatic surface, which is represented internally by the concave appearance of the septal surface (Fig. 4a, b). Absence of the atrioventricular part of the inlet ventricular septum is shown by three abnor-

Fig. 4 (a) Left ventricular view of a small AVSD (the same heart as in Fig. 1). The left valve is attached to the rim of the outlet septum. The defect of the inlet septum (transilluminated), although quite small, is large enough to cause the abnormal orientation and attachment of the valve leaflets. PA, pulmonary artery. (b) Left ventricular view of large AVSD. There is no inlet portion and the outlet septum is very attenuated. The inlet deficit extends above, below, and behind the left valve. The anterior leaflet of the valve is attached by secondary chordae tendineae to the rim of the outlet septum, while those of the septal leaflet insert into the summit of the remaining septum. Other abbreviations as in Fig. 1.

malities. These are the abnormal plane of the atrioventricular valve(s), which are attached at the same point to the septum (Fig. 1a, b), the loss of anteroposterior dimension to the left ventricular outflow tract (Fig. 2a, b), and the abnormal position of the aortic valve. This structure is no longer wedged between the ventricular inlets, but lies anterior and superior to the atrioventricular valve leaflets (Fig. 2b, 4b). All these features indicate defective development of the inlet septum and absence of its atrioventricular component. Specimens without an interatrial communication ("canal-type" ventricular septal defects) have in the past been excluded from the compass of the malformation under review³² but, if there is a disproportion between the inlet and the outlet lengths of the left ventricle, they can be classified as examples of atrioventricular septal defect.⁷

(2) ATRIOVENTRICULAR VALVES

Because the valves develop from the posterior wall of the embryonic heart and some elements of the inlet/

outlet cushions they are obligatorily malformed when the embryonic structures develop abnormally. Their myocardial origin is often suggested by the muscular anomalies of both chordae tendineae and papillary muscles in atrioventricular septal defect (Fig. 5). Valve morphology accords with the degree of posterior septum deficit—two orifices usually occur with a fairly small defect and the severest form of the malformation has the free floating leaflets of the common valve sometimes predominantly opening into one or other ventricle and the huge defect both behind, below, and in front of the common valve (Fig. 4b). The plane of the valves or valve, always abnormal in atrioventricular septal defect, is governed in part by the size of the remnant of the inlet septum. Usually the valves are attached to this structure so that they "descend" to an undue extent into the ventricles. Thus there is a gap where the septum should be, and interatrial or ventriculoatrial shunting occurs through this gap.

Ventriculoatrial shunting may also occur through

Fig. 5 The right valve in AVSD, right ventricular view. The valve leaflets are supported mainly by muscle. There are truly tendinous cords only at the right lateral-posterior commissure (arrowed). Most of the remaining tensor apparatus is muscular. P, pulmonary valve; RV, right ventricle; PL, posterior leaflet; ARL, anterior right leaflet.

the “cleft” in the left valve. The disposition of leaflet tissue is abnormal in atrioventricular septal defect so that the left valve leaflets are abnormally orientated. The anterior leaflet itself is not cloven but is always intact—the cleft is the tissue deficit between the anterior and posterior (septal) leaflets of the valve (Fig. 6a, b). The degree of deficit, however, does not necessarily influence the function of the valve.³⁵

Double orifice left atrioventricular valve results from an extra deep scallop (cleft) between either the anterior and lateral or the lateral and posterior leaflets, respectively, and a bridge of tissue between the two leaflets. Deep scallops (clefts) are sometimes found in normal hearts, especially in the tricuspid valve,³⁶ and reflect the undermining process which modelled them.

Parachute mitral valve, present in about 7% of cases,³⁷ results from inadequate liberation of the future papillary muscles from the posteromedial embryonic wall.

(3) AORTIC OUTFLOW TRACT

The pathognomonic appearance of the left ventricular outflow tract results from posterior septal deficiency (the “scooped-out” appearance)^{5,6} and the abnormal attachment, often with short, thick chordae,³⁸ of the anterior left leaflet to the remaining outlet septum. Because this septum does not have its normal smooth continuity with the infundibular septum, the myocardium in the outflow tract attenuates, giving the characteristic tunnel-like outflow tract (Fig. 4b).

The displacement of the aortic valve is probably real; if the atrioventricular septum is absent, then the aortic valve is further superior and to the right than in normal hearts, as it is no longer wedged between the two atrioventricular valves.

(4) PULMONARY OUTFLOW TRACT

Fallot's tetralogy and atrioventricular septal defect are an uncommon combination; the hypothesis offered concerning the inlet septal maldevelopment does not contradict the generally accepted views about the ontogeny of the infundibular malformation.^{33,34}

(5) ATRIAL SEPTUM

The atrial septum is often normally developed in atrioventricular septal defect, and this also lends support to the view that the malformation concerns the ventricular septum. None of the widely accepted views about atrial septation is challenged—rather, these views are supported. Defective atrial septation has never satisfactorily explained atrioventricular septal defect. Despite the usual cognomen “atrioventricular canal” the malformation does not remotely resemble the arrangement in the newly-looped embryonic heart. Arrested or abnormal development of this region cannot explain the malformation of the outflow tract of the left ventricle, which may be mild or severe, but is always present.

Common atrium does, of course, represent rudimentary atrial septation, and is usually associated

Fig. 6 (a) The left ventricular inflow in the normal heart. The anterior mitral leaflet is supported by both papillary muscles; the posterior leaflet is characteristically scalloped (1, 2, 3). The anterior leaflet completely covers the upper third of the ventricular septum and leaflet tissue is continuous around the annulus. (b) Left ventricular inflow in AVSD. The anterior leaflet of the left valve is almost at right angles to the normal structure in (a) so that the upper third of the ventricular septum is visible. The leaflet is supported only by the anterior papillary muscle group—the remainder of the leaflet inserts into the posterior wall of the outlet septum. Distinct septal and lateral leaflets are recognised. Although there is valve tissue on the summit of the ventricular septum there is a gap (the cleft, arrowed) between the anterior and septal leaflets. The anterior leaflet is never split—clefts are the spaces between the leaflets. PPM, posterior papillary muscle group; APM, anterior papillary muscle group. Other abbreviations as in Fig. 1.

with the more severe forms of atrioventricular septal defect (Fig. 6). There seems no reason to suppose any more elaborate mechanism than defective atrial septation to explain this coexistence.

Conclusion

The embryological studies reviewed here shed light on the developmental background of the ventricular septum and the atrioventricular valves. The embryology of the anatomical touchstones of atrioventricular septal defect has been hypothesised, and this hypothesis challenges neither the accepted views about atrial septation nor about outflow tract maldevelopment which may coexist.

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