Common or Single Ventricle

An Angiocardiographic and Hemodynamic Study of 42 Patients


SUMMARY To correlate anatomy with hemodynamics, the angiocardiographic findings were reviewed in 42 patients with common ventricle (CV). Nine had normally related great arteries (NRGA), 12 d-malposition, 21 l-malposition and 5 a common atrioventricular valve. Selective outlet chamber (OLC) angiocardiograms were available in 14 out of 29 patients with OLCS.

OLC position varied from anterior and to the right of the CV to posterior and to the left of it; two categories (anterior and lateral OLC) were delineated by a line 45° to the left of anterior in the horizontal plane. The OLC was anterior in all patients with NRGA, lateral in most l-malpositions, and almost equally divided between anterior and lateral in d-malposition (P < 0.05).

Complete hemodynamic data were obtained in 29 patients. Complete mixing of venous return occurred in four patients with atresia of one valve. In the remainder complete mixing occurred in 36%, unfavorable streaming in 12% and favorable streaming in 52%. Semilunar valve position and pulmonary stenosis did not affect the nature of mixing. Systemic arterial (SA) minus pulmonary arterial (PA) O₂ saturation was positive and significantly higher in patients with malposition with lateral OLCs than anterior OLCs (P < 0.001). However 79% of SA O₂ saturation variation could be predicted from pulmonary and systemic blood flow alone.

COMMON OR SINGLE VENTRICLE presents to the investigating cardiologist perhaps the most challenging diagnostic problem in congenital heart disease. This was so even before there was any prospect of surgical correction of the lesion. Now that reports of operative repair of this anomaly are emerging, and yet no clear criteria for selection of patients are established, it is evident that far more detailed preoperative investigation is now needed. A preliminary review of angiocardiograms of common ventricle have demonstrated some significant differences from other published reports. This prompted us to analyze cardiac investigation data in greater detail and to see to what extent the hemodynamic state could be explained by the angiocardiographic anatomy, and to what extent it was influenced by other factors.

Definitions and Classification

The problems of matching angiocardiographic with pathologic classifications of common ventricle have previously been discussed. Our classification is based upon that of Hallermann et al., modified in the light of pathological studies by Quero, and van Praagh et al., and our own radiological experience of selective angiocardiography of hypoplastic ventricles in mitral and tricuspid atresia.

Common or single ventricle is that condition in which both atrioventricular valves, or a common atrioventricular valve, are anatomically related to, and communicate actually or potentially with, a single ventricular chamber. If an outlet chamber is present, neither atrioventricular valve annulus is anatomically related to, or communicates actually or potentially with, this chamber.

This definition therefore encompasses single (primitive) ventricle, common ventricle, and those forms of mitral and tricuspid atresia in which the atretic valve is anatomically related to and therefore communicates potentially with the same common ventricular chamber as the patent atrioventricular valve.

Malposition of the great arteries exists when the semilunar valves and great arterial trunks are abnormally unrelated in space. In d- and l-malposition the midpoint of the aortic valve lies respectively to the right and left of the midpoint of the pulmonary valve.

It is recognized that when the aorta arises from the outlet chamber and the pulmonary artery from the common ventricle that transposition exists, if in fact the bulboventricular septum, which lies between the common ventricle and outlet chamber, is the true interventricular septum. However, un-
les oblique projections to profile the bulboventricular septum are filmed it may not always be possible angiocardio-
graphically to state with certainty whether one or both great
arteries originate from the outlet chamber or the common
ventricle, so we have preferred for practical reasons not to
distinguish between malposition and transposition.

The mitral and tricuspid valves, for the purposes of this
communication are termed left and right atrioventricular
valves, since we have found it impossible to distinguish the
morphology of these valves by angiocardiology, and
acknowledged authorities have found this to be difficult even
at autopsy. Furthermore, in contrast to classical corrected
transposition we have found it impossible on angiocardio-
graphic grounds to deduce accurately the direction of
bulboventricular looping. Since all but one patient with two
atrioventricular valves had visceroatrial situs solitus, in all
but this patient the left atrioventricular valve was related to
the morphological left atrium.

A common atrioventricular valve has the anatomic
appearances of the common valve in complete atrio-
ventricular canal, and in particular is invariably associated
with an ostium primum atrial septal defect.

Double outlet bulbus exists when both great arteries
originate from the outlet chamber. This is invariably
associated with a bilateral conus.

Bilateral conus exists when neither atrioventricular valve
is in continuity with either semilunar valve. This may occur
in the absence of an outlet chamber, so it is not synonymous
with double outlet bulbus.

Because a common atrioventricular valve in the presence
of common ventricle is so frequently associated with com-
plex abnormalities of cardiac position, situs and venous con-
nection, patients in this category are dealt with separately
from the rest, who are divided into those with l-malposition,
d-malposition and normally related great arteries.

Material and Methods

The case material consists of all patients studied over the
last five years at Killingbeck Hospital in whom a positive
angiocardiographic diagnosis of common ventricle could be
made. The age at investigation ranged from 1 day to 37
years (median 3 years). There were 22 females and 20 males.
Intracardiac operation was performed in nine patients, four
of whom subsequently were autopsied. A further nine autops-
ies were available for review. Thus independent confirm-
ation of the diagnosis was present in four of eight patients
with d-malposition, five of 20 patients with l-malposition, seven of nine patients with normally related great arteries
(one of whom had left atrioventricular valve atresia), and in
two of five patients with a common atrioventricular valve.
Of these last five, one had dextrocardia with bilateral four-
lobed lungs and d-malposition, one had levocardia in situs
inversus with d-malposition, one had dextrocardia in situs
solitus with d-malposition, one levocardia with bilateral
right lung and d-malposition and one levocardia with
bilateral left lung and l-malposition. Thoracic situs was
assessed in vivo in four of these patients by bronchial
tomography.

Patients were investigated breathing room air, sedated
with 0.1 ml/kg of a mixture containing 25 mg chlorproma-
zine, 6.25 mg promethazine and 6.25 mg meperidine per 1
ml.

Simultaneous straight biplane angiocardiograms in the
frontal and lateral projections were performed in all but one
patient (who had at surgery normally related great arteries
and no outlet chamber), in whom a left anterior oblique
cineangiocardiogram was utilized. The majority of films
were taken with an Elema-Schönander roll-film changer,
but over the past year Siemens biplane cineangiography
using a 6 or 10 inch field has been increasingly used. Selective
injections into the common ventricular chamber were
invariably performed with the catheter through either atrio-
ventricular valve or both in turn, using the largest NIH
catheter that could be introduced (never less than 6F and
usually 8-10F) and a dose of 1.7-1.8 ml/kg of 70% Conray
(sodium iothalamate). In the patients with right or left atrio-
ventricular valve atresia, injections were made respectively
into the right and left atria. Rather smaller doses of contrast
medium were used for selective outlet chamber injections,
which were performed with the catheter tip sufficiently far
into the chamber to ensure that it was completely opacified
in each case.

Identification of atrioventricular valves was tabulated ac-
cording to two categories, direct and indirect visualization.

Direct visualization indicates either (a) that the valve
through which the catheter was placed was seen to close
around it during systole, and that nonopacified blood was
seen to traverse the valve during diastole or (b) that a por-
tion of the valve annulus was identified and nonopacified
blood was seen to traverse it.

On injection of contrast medium into the common ventri-
cle both annulus and nonopacified blood are visualized dur-
ing diastole but not systole. The annulus is relatively im-
mobile and has a sharply defined crescentic or elliptic
margin, whereas the interface between nonopacified blood
and contrast medium is diffuse and the valve leaflets, when
seen, are more sharply defined but mobile.

Additional confirmation of identification of the right
atrioventricular valve annulus is that in the lateral projection
it is seen to be closely related to the right coronary artery.
The left circumflex coronary artery is closely related to the
lateral margin of the left atrioventricular valve annulus.

Indirect visualization of an atrioventricular valve indi-
cates one of two things. Either nonopacified blood (but not
the annulus) was seen entering the ventricle in a site differ-
ent from the directly visualized valve through which the catheter
was introduced or, more commonly, after injection of con-
trast medium with the catheter through the right atrio-
ventricular valve the left atrioventricular valve was
visualized after pulmonary venous return in a different posi-
tion from the right sided valve, but entering the same ven-
tricle. The diagnosis of common atrioventricular valve was
only accepted without autopsy if the following criteria were
fulfilled. (a) Two atrial chambers could not be distinguished
manometrically, angiocardiographically, or oximetrically.
(b) Contrast medium passed through the same atrio-
ventricular valve from systemic veins as from pulmonary
veins. (c) Only one atrioventricular valve annulus was
visualized on injection into the common ventricle and none.

*This includes one patient with d-malposition in situs inversus, treated
throughout as an l-malposition.
on injection into the outlet chamber (if present). (d) One atrioventricular valve was demonstrated by angiocardiography.

The presence or absence of pulmonary stenosis and an outlet chamber was assessed separately by observers not aware of the hemodynamic and autopsy findings. An attempt was made at visualizing papillary muscles, and the trabecular pattern of the common ventricle towards end-diastole was also analyzed so as to classify this into the two categories left ventricular type and primitive type.  

The position of the outlet chamber relative to the common ventricle was assessed by comparisons of separate injections into the two chambers. Relative positions were assessed in the region in which the two chambers were contiguous, i.e., in the region of the bulboventricular septum and foramen, and recorded as if on a standard horizontal plane vectorcardiographic display. Where a selective outlet chamber injection was not available rather more reliance was placed on the orientation of the bulboventricular septum. Outlet chamber position was assessed to the nearest 30°. Thus a position of 0° (fig. 6) indicates that the bulboventricular septum is profiled in the frontal projection and the long axis of the leftward outlet chamber is more or less vertical. +90° indicates an anterior outlet chamber with its long axis more or less vertical and the bulboventricular septum profiled in the lateral plane. +60° indicates that the bulboventricular septum is not profiled in either plane, but that the outlet chamber is more anterior than leftward.

If angiocardiography preceded definitive hemodynamic investigation, no measurements were made until at least 30 minutes after angiocardiography. The pulmonary artery was usually entered with a standard NIH catheter, though in two patients this was passed from the aorta, via a patent ductus arteriosus in one and a surgical aorta/right pulmonary anastomosis in the other. In two patients a Swan-Ganz balloon catheter was used and in four patients with malposed great arteries a Shirey catheter was passed from an axillary arteriotomy retrogradely through the aortic valve, looped in the common ventricle and made to cross the pulmonary valve. The same technique was used to enter the left atrium retrogradely through the mitral valve in four patients. Femoral artery pressure was usually monitored with an indwelling needle or cannula throughout the investigation, but in some infants was introduced only when the pulmonary artery was first entered.

Once the catheter was in the pulmonary artery, simultaneous femoral and pulmonary artery pressures were recorded and blood samples taken for oximetry and blood gas determination. While three 45 second collections of gas were made to determine O₂ uptake by the flow-through method, the catheter was withdrawn and pressures and samples for oximetry and blood gas determination rapidly obtained from superior vena cava (SVC), inferior vena cava (IVC) and, where possible, from the left atrium and pulmonary veins. Samples for oximetry were taken at least in duplicate except in small infants, and analyzed with a Kipp M01 hemofluxor, which as used in our laboratory has 95% reproducibility limits of ± 2.5%. Pulmonary (Qp) and systemic (Qs) blood flow were determined by the Fick technique, allowing 0.3 ml dissolved oxygen in the plasma per 100 ml/100 mm Hg, and using the formula mixed venous O₂ saturation = (3 × SVC saturation + IVC saturation) ÷ 4.  

Pulmonary venous saturation, when unknown, was assumed to be 97%. In those patients studied prior to the development of the method for measurement of Qs consumption, values were predicted from age, height and weight. Indocyanine green curves were recorded using a Waters XC302 curve and D400 densitometer.

Analysis of enumeration statistics was carried out where possible using the Chi-square test. Otherwise complex tables were broken down into fourfold tables, each of which was analyzed by the exact fourfold test, for which two-tailed probabilities are quoted. For analyzing continuous variables, extensive use was made of stepwise regression analysis, using a program based upon the logic of Efron's modified to suit a 4K digital computer (Linc 8, Digital Equipment Corp.). The dependent variable chosen was systemic arterial O₂ saturation. The independent variables introduced to predict SA O₂ saturation were Qp, Qs, pulmonary resistance (Rp), systemic resistance (Rs), log Rp, Qp/Qs, Rp/Rs, hemoglobin concentration and ventricle to PA peak systolic gradient. Dummy variables were used to separate patients into various categories, namely (a) with and without pulmonary stenosis, (b) with and without outlet chambers, (c) d-malposition, l-malposition and normal relation of the great arteries, (d) with and without measured O₂ consumption, (e) with and without previous palliative surgery, (f) position of outlet chamber (see below), and (g) presence or absence of valve atresia. On receiving this information, the computer then selects that dependent variable (say A) which correlates best with SA O₂ saturation. The effect of this variable on the prediction is then eliminated, and from the remaining independent variables the most highly correlated (say B) with SA O₂ saturation is again selected. After a third variable (say C) has been selected it may be that C in combination with A (or B) may predict the SA O₂ saturation so well that B (or A) may be rejected. This procedure is automatically carried out by the computer after each new variable is entered into the predictive equation. The level for selection or rejection of a dependent variable was set at an F ratio of 2 which, with the numbers involved, would in practice mean that failure to select a variable would strongly indicate that it was nonpredictive. Deviations from the final predictive equations were then studied for evidence of nonlinearity or correlation with variables not entered.

**Results**

**Identification of Atrioventricular Valves (fig. 1)**

Of the 34 patients with two atrioventricular valves, the right atrioventricular valve was identified directly in 33 (97%), and the left atrioventricular valve directly in 28 (82%). Simultaneous direct visualization of both valves was obtained in 20 (59%) and in half of these the two contiguous valves gave rise to a characteristic 'figure of 8' sign in either the lateral or frontal projection. In the remainder the atrioventricular valves were visualized indirectly.

Two patients had left atrioventricular valve atresia with normal relation of the great arteries, with the atresia documented by selective left atrial angiocardiography (fig. 2). In both of these the right atrioventricular valve was
directly visualized and the atretic left atrioventricular valve was visualized bulging into the common ventricle during ventricular diastole with its annulus outlined.

One patient had right atrioventricular valve atresia with 1-malposition and in this patient the left atrioventricular valve was directly visualized. On selective outlet chamber injection no atretic valve was seen (fig. 3).

Five patients had a common atrioventricular valve. In three of these a diastolic goose-neck deformity was present (fig. 4A) and in a fourth (fig. 4B) a cleft leaflet was observed in systole.

**Position and Anatomy of the Outlet Chamber**

An outlet chamber was identified in 29 patients (69%) and selective outlet chamber angiocardiography was performed in 15 (52%) of these. Angiocardiography predicted correctly the presence or absence of an outlet chamber in all the 18 patients operated on or autopsied. No significant association was demonstrated between the presence or absence of an outlet chamber and great vessel relationships or presence of a common atrioventricular valve.

The base of the outlet chamber invariably was related to the aorta when the great arteries were malposed and to the

**FIGURE 1.** A) Common ventricle without outlet chamber, 1-malposition, pulmonary stenosis, and dextrocardia with situs solitus. Diastolic frame (frontal projection) shows both atrioventricular valves in continuity with one another (black arrows) forming most of a figure of 8. The catheter is through the tricuspid valve. The trabeculations are of left ventricular type. B) Common ventricle without outlet chamber, d-malposition, mild pulmonary stenosis and severe pulmonary vascular disease (lateral projection). The catheter is through the right atrioventricular valve (r) and the left atrioventricular valve annulus (l) is visualized adjacent to, but behind and above it. There is a bilateral conus, and the conus septum (arrow) is prominent. The trabeculation is characteristic of neither right nor left ventricle, and is therefore termed primitive in type.

**FIGURE 2.** Common ventricle with left atrioventricular valve atresia, normal relation of great arteries and subpulmonary outlet chamber. Lateral view, common ventricular injection, selected for demonstration of bulboventricular foramen (solid white arrow) and outlet chamber (black arrow), rather than left atrioventricular valve, which is however seen as a crescentic indentation of the upper posterior ventricular cavity (open white arrow). Note the normal sized aorta, and the pulmonary artery band. Left atrioventricular valve atresia was confirmed during atrial septectomy. A 24 mm Hg mean pressure gradient was present between the two atria, the left being higher. The patient had undergone pulmonary artery banding five years previously. A left atrial angiogram showed a normal sized left atrium, mitral atresia and opacification of the right atrium alone initially, through a patent foramen ovale.
Figure 3. Common ventricle with right atrioventricular valve atresia, l-malposition, subaortic outlet chamber, and moderate pulmonary valve stenosis. The right atrial angiocardiogram showed appearances typical of tricuspid atresia. A) Retrograde injection into common ventricular chamber (frontal projection). The posterior left atrioventricular valve annulus is faintly seen (black arrows). There is a subaortic outlet chamber on the left upper shoulder of the heart. The catheter demonstrates the position of the aorta, which has not yet opacified. The trabeculations of the common ventricle are left ventricular in type. B) Retrograde injection into outlet chamber (lateral projection). Neither in this nor the frontal projection was any valve identified in the outlet chamber. The left atrioventricular valve is visualized posteriorly in the common ventricle, opacified via the bulboventricular foramen. Note the heavy trabeculation. C) For comparison, the lateral projection of a selective right ventricular cine angiocardiogram in a patient with tricuspid atresia, transposition of the great arteries (S, D, L146) and pre-ductal coarctation. The looped catheter has its tip in the pulmonary artery. The atretic tricuspid valve (arrow) relates to the hypoplastic right ventricular sinus as confirmed at autopsy, when chordae tendineae were found inserted into this valve. Common ventricle is not present in this patient.

Figure 4. A) Common ventricle with common atrioventricular valve, l-malposition, subaortic outlet chamber, pulmonary atresia, total anomalous pulmonary venous drainage to left SVC, dextrocardia and thoracic isomerism (bilateral 4-lobed lungs; splenic status uncertain). Autopsy confirmation. The common atrioventricular valve (frontal projection) is large and extends superiorly almost to the superior wall of the common ventricle giving an appearance similar to the classical gooseneck deformity of complete atrioventricular canal. There is a subaortic outlet chamber (o) which was more clearly demonstrated by selective injection into it. B) Common ventricle without outlet chamber with common atrioventricular valve, l-malposition, bilateral conus, severe subvalvular and valvular pulmonary stenosis, right aortic arch, levocardia with bilateral left lung and interrupted inferior vena cava. The common atrioventricular valve shows a cleft in systole characteristic of endocardial cushion defects. Injection of contrast medium into the superior vena cava of this patient demonstrated opacification of the same atrium as opacified on pulmonary venous return.
pulmonary artery when the great vessels were normally related. Three patients had a double outlet bulbus.

The outlet chamber communicated with the common ventricle by way of the bulboventricular foramen and, in two patients, a more inferior defect (fig. 5A). The bulboventricular foramen frequently appeared to have a diameter less than that of the aortic root without there being hemodynamic evidence of obstruction, perhaps indicating that the foramen was elongated rather than circular (fig. 5B). Conversely in the only patient with an aortic outflow gradient of more than 10 mm Hg (a 60 mm gradient across the bulboventricular foramen) no angiographic stenosis was observed.

The apical recess of the outlet chamber (that portion between the bulboventricular foramen and its apex) varied in length from being less than the diameter of the aortic root, to extending the full length of one surface of the heart. No papillary muscles were seen in the outlet chambers of any patients with selective outlet chamber angiograms. However, papillary muscles were infrequently identified within the common ventricle. In no autopsied patient was any tensor apparatus identified within an outlet chamber.

The orientation of the outlet chamber was described in terms of its relationship to that part of the common ventricle with which it was contiguous. Because of its size and relationship to the great arteries it was invariably for the most part superior to the common ventricle, but its disposition in the horizontal plane varied from anterior and to the right to posterior and to the left of the common ventricle (figs. 5, 6). Two groups were delineated, separated by an arbitrary line drawn at +45°. Those with outlet chambers in an anterior, or principally anterior, position are classified as having an anterior outlet chamber. Outlet chambers principally in the lateral position are termed lateral outlet chambers. There was a significant (P < 0.05) association between orientation of the outlet chamber and position of the great arteries. Patients with normally related great arteries had anterior outlet chambers whereas those with l-malposition tended to have lateral outlet chambers. Outlet chambers in d-malposition were almost equally divided between the two groups.

**Trabecular Pattern of Common Ventricle**

In every case with an outlet chamber, that chamber was more heavily trabeculated than the common ventricle. In assessing the trabecular pattern of the common ventricle it became clear that though two ends of a spectrum could be readily differentiated, there was no clear cut boundary apparent between the primitive and the left ventricular type of trabeculation described by Marin-Garcia et al.³ The left ventricular type tended to be associated with l-malposition and normally related great arteries, while the primitive type was more associated with common atrioventricular valves.

**Angiocardiographic Appearances of Pulmonary Stenosis**

The angiocardiographic diagnosis of pulmonary stenosis was made 'blind' by one observer. In the 30 patients in
whom the pulmonary artery was entered at catheterization the ventricle to pulmonary artery systolic gradient was 30 mm Hg or more in 16 patients, one of whom had pulmonary atresia. In 5 of these (31%) the angiocardiographic diagnosis was not made. In one such false negative the valve gradient was 148 mm Hg. However, there were no false positive angiocardiographic diagnoses. In the 12 patients in whom the pulmonary artery was not entered there was severe stenosis angiocardiographically in eight, pulmonary atresia in two, and no angiocardiographic stenosis in two. In the latter two one cannot be sure whether pulmonary stenosis was in fact present, so the overall incidence of definite pulmonary stenosis or atresia was 26 out of 40 (65%). No significant association could be demonstrated between pulmonary stenosis or atresia and great vessel position. Four out of five patients with common atroventricular valve had severe pulmonary infundibular and valvular stenosis and the other had pulmonary atresia. This incidence was not significantly different from that in the remainder of patients.

Pulmonary outflow obstruction was present in three patients with normally related great arteries, two of whom had outlet chambers. In one of the latter, the obstruction was mild (30 mm Hg gradient) and at the site of the bulboventricular foramen. In the other two there was infundibular stenosis, the length of which was much greater than the subvalvular pulmonary stenosis characterized in patients with malposition, in whom the obstruction was much more localized, lying immediately beneath the pulmonary valve. Pulmonary stenosis in the malposition patients was valvular in all patients and also subvalvular in all but two, as assessed angiocardiographically.

Five out of six patients with a right aortic arch had pulmonary stenosis as opposed to 21 out of 36 patients with a left arch. These proportions do not differ significantly.

**Coronary Artery Anatomy**

The detailed anatomy of the coronary arterial system could be established from aortography or selective outlet chamber angiography in six patients. Five of these had l-malposition, and of these the anterior descending coronary artery originated from the right coronary artery in four and the left coronary artery (autopsy proven) in one. One patient had normally related great arteries and a normal coronary artery system.

**Hemodynamic Findings**

The pulmonary artery was entered in 30 patients (71%) and there were only two patients (studied very early in the series) without severe angiocardiographic pulmonary stenosis or atresia (<2 mm orifice) in whom the pulmonary artery was not entered. In one patient with severe pulmonary stenosis, entry of the catheter into the pulmonary artery provoked a sharp fall in systemic arterial oxygen saturation which resolved on withdrawal of the catheter. Because of the resultant hemodynamic instability this patient will not be considered further. Of the remaining 29, the left atrium and pulmonary veins were entered in 16 (55%). Two failures to enter the left atrium resulted from an intact atrial septum and severe mitral stenosis.

Of these 29 patients seven had had previous palliative surgery. Three (including one with left atroventricular valve atresia) had had banding of the pulmonary artery, one a barely functioning ascending aorta to main pulmonary anastomosis, two an aorta to right pulmonary artery anastomosis and one a subclavian/pulmonary artery anastomosis. Of these three one had congenital pulmonary atresia and two virtual pulmonary atresia even before palliation, so it is unlikely that in any palliated patient there was a significant difference in oxygen saturation in the two pulmonary arteries.

**Comparison of Systemic (SA) and Pulmonary (PA) Arterial O₂ Saturations**

Inspection of figure 7 shows that patients with pulmonary or atroventricular valve atresia all had complete mixing, that their systemic and pulmonary arterial O₂ saturations were not significantly different. Of the remaining 25, nine (36%) had complete mixing, three (12%) had unfavorable streaming and 13 (52%) had favorable streaming.

In the group as a whole the most favorable streaming by far was in two patients who had had pulmonary artery banding, one with l-malposition and one with normally related great arteries.

The mean O₂ saturation difference (SA-PA) in those two was 19.9% (SE = 4.8%) whereas in the remaining 23, the mean difference was 2.4% (SE = 1.11%). The means are highly significantly different from each other (P < 0.001) and the mean in the nonbanded group is significantly different from zero (P < 0.05). Subdividing the nonbanded 23 into those with l-malposition, d-malposition and nor...
mally related great arteries revealed no significant difference between the groups; nor did subdividing into those with and without previous shunts, or those with and without pulmonary stenosis. The proportion of patients with complete mixing was not significantly different in those with and without pulmonary stenosis and no correlation was apparent between Qp and SA-PA in the group as a whole, or those with or without pulmonary stenosis.

However, when those patients with malposition, without previous surgery, and with two atrioventricular valves were subdivided according to the position of the outlet chamber, the mean difference in those with an anterior outlet chamber was $-0.25\%$ ($SE = 1.89\%$) whereas in those with a lateral outlet chamber, it was $4.56\%$ ($SE = 0.67\%$). This difference between the two groups is highly significant ($P < 0.001$) (fig. 8), and it is only the lateral outlet chamber group which is significantly different from zero ($P < 0.001$).

**Determinants of Systemic Arterial Oxygen Saturation**

The results in this section relate almost entirely to stepwise regression analysis.

There were 29 patients in whom stable hemodynamic conditions were present. When analyzing the group as a whole in terms of those variables which predict systemic arterial $O_2$ saturation best (namely Qp and Qs), introduction of (a) measured vs predicted $O_2$ uptake, (b) previous palliative operation vs no previous palliative operation, and (c) valve atresia vs no valve atresia indicated that these different categories were nonpredictive. They therefore could be pooled without introducing any systemic error.

However, patients with measured $O_2$ uptake were initially separated from the group as a whole in order to see whether the use of predicted $O_2$ uptake was associated with a larger random error of prediction.

In the group with measured $O_2$ uptake (19 patients), when Qp/Qs was introduced together with Qp and Qs, Qp/Qs was selected first ($P < 0.001$), then Qs ($P < 0.025$), then Qp ($P < 0.05$), and finally Qp/Qs was rejected again, indicating that when Qp and Qs are given, Qp/Qs had no additional predictive value. If Qp/Qs was not introduced Qp was selected first ($F_{1,17} = 16.9, P < 0.001$) and then Qs ($F_{1,16} = 22.1, P < 0.001$). The complete predictive equation was:

$$FA \ O_2 \ saturation = 84.5 - 4.78 \ Qs + 1.91 \ Qp$$

This explained 79% of the variation in systemic arterial $O_2$ saturation observed and the residual standard deviation was 5.9%. The standard errors of the regression coefficients were 0.345 for Qp and 1.017 for Qs.

The results in the combined group (with and without measured $O_2$ consumption) were qualitatively and directionally identical, but despite the larger numbers (29) involved, only 64% of the variation was explained by Qp and Qs and the residual standard deviation was 7.3%.

The prediction was not significantly different for those patients with and without pulmonary stenosis. These two groups were analyzed separately, excluding banded patients so as to look at the nature of the effects of pulmonary resistance and pulmonary stenosis. A 41 mm Hg boundary for pulmonary outflow systolic gradient was drawn between these two groups because one patient with a gradient of 40 mm Hg had an Rp of 15.5 units $\cdot m^2$ without stenosis and it was felt that this would be the principal determinant of pulmonary flow. A further patient with a gradient of 148 mm Hg across the pulmonary valve, and a 60 mm Hg gradient across the bulboventricular foramen leading to a subaortic outflow chamber was eliminated from both groups because of the effect of the latter on resistance to aortic outflow.

This left 13 patients with pulmonary stenosis. As expected, Qp and Qs were both predictive, but when these were not used, ventricular/pulmonary artery systolic gradient was not predictive even in combination with Rs and Rp.

In the 14 patients without pulmonary stenosis, Rp/Rs was predictive though less so than Qp/Qs ($r = -0.60$, $F_{1,12} = 6.67$). Neither Rp nor Rs added significantly to this prediction though there was a negative correlation between...
Rp and SA O₂ saturation ($r = -0.55$). The equation relating FA O₂ saturation and Rp/Rs was:

$$\text{FA O}_2 \text{ saturation} = 89.0 - 15.8 \text{ Rp}/\text{Rs. (Sy. x = 7.16%).}$$

The relationship between age and Rp is demonstrated in figure 9. Excluding two patients with stenosis, the data were best fitted with a curvilinear plot. The 14 patients with pulmonary stenosis had a significantly lower ($P < 0.05$) mean systemic arterial O₂ saturation (75.6 ± 3.3%) than those eight patients without pulmonary stenosis or elevation of pulmonary resistance above 10 units m⁻² (85.0 ± 2.4%).

Despite the relationship between outlet chamber position and favorable streaming already demonstrated, when this was introduced as a dummy variable, at no point did it prove predictive after Qp and Qs had been given: the same was true for great vessel position, and Hb concentration as a continuous variable.

In four patients in all (9.5%) the systemic arterial O₂ saturation was >90%, and in three of these indocyanine green dye curves were originally reported as showing no right-to-left shunt on injection into the SVC and IVC, though there was early appearing dye on injection into the ventricle. Only on review of the dye curves was it apparent that a very slight, and certainly unquantifiable right-to-left shunt was present. And in one of these patients the mean of four arterial oxygen saturations breathing room air was 97.2% ($P_O = 86 \text{ mm Hg}$). This patient also had complete heart block.

### Discussion

#### Morphology

In classifying common or single ventricle with outlet chamber, it has been customary to subdivide according to the position of the outlet chamber; that is to say whether it lies to the right or left of the common ventricle. When it lies to the right the condition has been variously described as noninversion of the infundibulum,¹⁹ and d-bulboventricular loop,²⁰ whereas when it lies to the left there has been said to be inversion of the infundibulum¹⁸ or an l-bulboventricular loop.²⁰ Our results did not enable any simple distinction of this kind to be made.

The discrepancy between our own angiocardiographic findings and those in two undoubtedly authoritative pathologic studies may be partly a question of definition. Angiocardiographically the cavity of the outlet chamber is demonstrated, whereas pathologically the delimiting coronary arteries also define its position. But the discrepancy in our view is principally a matter of whether the heart is studied inside or outside the body. If the diagram of the angiocardiographic position of the outlet chamber (fig. 6) is rotated 45° in a clockwise direction, then what we have termed an anterior outlet chamber would correspond roughly to d-bulboventricular looping and what we have termed a lateral outlet chamber would correspond to l-bulboventricular looping. This interpretation is supported by the observation that with normally related great arteries (where d-looping has almost certainly occurred) the outlet chamber was invariably anterior whereas in l-malposition (where there has probably been l-looping) it was usually lateral. The same explanation can be put forward for the fact that angiocardiographically outlet chambers directed leftward and posteriorly have been described not only here, but also briefly elsewhere whereas this position of the outlet chamber is not mentioned in several large pathologic series.⁹,¹³,¹⁹-²²

The approach of Quero to the question of atresia of one atrioventricular valve in single ventricle,⁶,⁷ recently espoused by Cabrera and colleagues,²³ depends upon the concept of the 'topographically homologous ventricle.' This in turn depends upon accurate deductions about bulboventricular looping. These are more reliable at autopsy than angiocardiography, since the trabecular pattern of the bulboventricular septum can be assessed as well as its orientation.

Because of (a) the problems of assessing trabecular pattern encountered, (b) the variety of outlet chamber positions and therefore orientations of the bulboventricular septum found, and (c) the overlap in all these variables...
between d- and l-malpositions, it was not felt feasible to deduce reliably the direction of looping from angiocardiographic appearances in many individuals. Previous pathological reports have indicated the problems of predicting bulboventricular looping from semilunar and great artery relationships.13, 20

The approach suggested here is therefore to concentrate simply on the relationship of the atretic valve to the ventricular chamber(s), and is supported by other pathologic studies24, 25 (Anderson, R.H., personal communication). If contrast medium is injected into the ventricular chamber immediately beneath the atretic left atrioventricular (mitral) valve, that atretic valve appears as a nonpatent bulge into the upper posterior part of the ventricular chamber as seen in the lateral projection. This has been confirmed at autopsy or operation in six patients with mitral atresia.8 If this bulge relates to the same ventricular chamber as the right atrioventricular valve then there is a common ventricle. In the usual form of mitral atresia without common ventricle, the bulge relates to a hypoplastic, smooth walled, directly posterior, left ventricle.8 However, in the two patients presented here a directly anterior subpulmonary outlet chamber was demonstrated which could not conceivably have been in relation with the left atrioventricular valve. Identical appearances were discovered on reviewing the angiocardiograms of a case previously reported by one of us;26 here the autopsy findings resembled closely those of 'atresia of the left atrioventricular orifice associated with a Holmes heart.'19

The situation in right atrioventricular (tricuspid) atresia is slightly different in that we have been unable to demonstrate the atretic valve related to the common ventricular chamber. Our own observations in patients with right atrial angiocardiograms typical of tricuspid atresia are that when contrast medium is injected into the small outlet chamber, the presence or absence of an atrioventricular valve within it can be detected with considerable accuracy. The presence of such a valve (as in fig. 3C) identifies the 'outlet chamber' as a hypoplastic right ventricle and rules out the diagnosis of common ventricle. The absence of such a valve is highly suggestive of common ventricle, particularly if (as in figs. 3A and B) the outlet chamber is relatively remote from the right atrium. The angiocardiographic anatomy of this patient corresponds with the pathologic description of Quero,8 except that in his case there was d-malposition, as opposed to l-malposition in ours.

Several distinguished authorities, while differing on other aspects of the definition of single ventricle, now agree that mitral and tricuspid atresia should be excluded from that definition.8, 13, 20, 22 The theoretical argument against this position has been admirably stated by Quero.5, 6

Briefly, three major hypotheses have been advanced as to the morphogenesis of common ventricle: (a) absence of the posterior interventricular septum,26, 27 (b) failure of rightward migration of the atrioventricular canal28 and (c) absence of one or both ventricular sinuses, or the sinus portion of the ventricular septum.26 The bulboventricular septum, (as we have termed it largely in order to differentiate it from a normal interventricular septum) is interpreted as true, but displaced interventricular septum according to (b)26 and (c),20 but not to (a).26 Examination of the specialized conducting tissue of the heart has not, as was hoped,9, 20 resolved this conflict.29 It should be emphasized, however, that not one of these theories, in itself, excludes atrioventricular valve atresia from common ventricle on morphogenetic grounds. Each is, instead, principally concerned with explaining the abnormal relationship in common ventricle between the bulboventricular septum and the atrioventricular canal. That this relationship can profoundly influence the relative size of the two cardiac chambers of bulboventricular origin has been elegantly demonstrated by experimental embryology with respect to both the right27, 20 and left31 atrioventricular valves. The size of the atrioventricular orifice and the extent to which it straddles the interventricular septum modifies the size of the ventricular chambers by altering the fetal blood flow into them.

This concept has interesting theoretical applications in the case of left atrioventricular valve atresia particularly when the great vessels are normally related. One would predict that if the interventricular septum were normally related to the atrioventricular canal, albeit with an associated ventricular septal defect, that the left ventricle would be hypoplastic along with the aortic valve and ascending aorta. This indeed happens in typical mitral atresia. By contrast when there is a common ventricle, the bulboventricular septum if present lies largely anterior to the atrioventricular canal and blood flow into the aorta is unobstructed. It is hardly surprising that in neither of our cases with common ventricle, and in none of those previously described5, 19, 20, 32, 33 was there hypoplasia of the aortic valve or ascending aorta. To sum up, in typical mitral atresia there is a large anterior (right ventricular) chamber and a small posterior (left ventricular) chamber, whereas when there is a common ventricle with outlet chamber, the position of the large and small chambers is reversed. This difference is so striking that it seems to us misleading not to distinguish mitral atresia with common ventricle from mitral atresia without it. When the position of atretic atrioventricular valves could not be established by angiocardiography, there were solid practical reasons for excluding atrioventricular valve atresias from common ventricle. In our view, the demonstration that it is possible angiocardiographically to deduce or identify directly the position of the atretic valve and its relationship to the main ventricular chamber justifies a reappraisal at this time.

Common atrioventricular valve with common ventricle is not an easy diagnosis to make in life. Though radiological descriptions of the entity are not lacking,5, 6, 21, 34 specific criteria for the diagnosis are lacking. Those we adopted may well have excluded some patients with a relatively small ostium primum atrial septal defect. Nevertheless, it is of some interest that the common atrioventricular valve showed some but not all features of complete atrioventricular canal in all but one case. The anterior common leaflet was undivided in both autopsied patients; when this is the case, the typical appearance of the left ventricular outflow tract may be absent.35

If surgery is planned, common ventricle with both atrioventricular valves patent must be distinguished from common atrioventricular valve and atrioventricular valve atresia. Hence our emphasis on direct as opposed to indirect visualization of the atrioventricular valves. In reviewing the
angiograms, it became clear that an unnecessarily large number of films during injection into the common ventricle had been taken. In 61% of cases, probably because of the large doses and rapid injection rates of contrast medium used, simultaneous direct visualization of both valves had been achieved with one common ventricular injection. More information could have been obtained had selective outlet chamber angiography, aortography, or oblique projections been filmed rather than repeat common ventricular injections with the catheter through the other atrioventricular valve. In particular, the figure of 8 appearance of two atrioventricular valves in continuity with each other seemed highly characteristic of common ventricle, though it was only seen in 30% of patients. At autopsy these valves are almost invariably adjacent.13, 32

The high incidence of subvalvular pulmonary stenosis in the patients studied also has important surgical implications, in view of the fact that in common ventricle with outlet chamber and malposition of the great arteries, the atrioventricular bundle has been shown to run anterior to the subpulmonary outflow tract.28 Since the right coronary artery which frequently gives origin to the anterior descending coronary artery also runs anterior to the pulmonary valve when there is l-malposition, this critical region is probably best left alone, and the pulmonary stenosis bypassed by some form of external conduit.1 Another important surgical consideration is whether the chordal apparatus of the two atrioventricular valves is separate or shared, but no way was found of answering this question angiographically.

Hemodynamics

Surprisingly little detailed information is published in the literature on the hemodynamics of common ventricle, particularly in the presence of pulmonary stenosis. In the four reports dealing with the largest number of living patients with common ventricle19, 31, 32, 34 the pulmonary artery was entered in only eight patients who had a gradient of 30 mm Hg or more across the pulmonary outflow. Rahimtoola and colleagues,36 showed that preferential streaming (i.e., incomplete mixing) occurred more often than not in common ventricle. Favorable streaming was more common with l- than with d-malposition. They also suggested that mixing would be more complete in the presence of severe pulmonary stenosis. Despite these findings, others have stated that 'the common ventricular chamber ensures a similar oxygen content of blood in each great vessel.27

The hemodynamic findings presented here support Rahimtoola's demonstration that complete mixing is the exception rather than the rule,36 but with respect to great vessel position and pulmonary stenosis differ significantly. No discernible effect of pulmonary stenosis upon the nature of the streaming was shown, and the pulmonary artery was entered in 15 patients with a pulmonary outflow gradient above 30 mm Hg. However pulmonary and atrioventricular valve atresia resulted in complete mixing. Pulmonary artery banding was associated with significantly more favorable streaming than in the remaining patients; the explanation for this observation is not known. There was no statistically significant difference between 1- and d-malposition in streaming; indeed the only other factor demonstrated to be associated with streaming was the position of the outlet chamber. The fact that patients with malposition and a lateral outlet chamber had significantly favorable streaming presumably resulted from (a) the fact that none had double outlet bulbus, i.e., all were transpositions with the aorta alone originating from the outlet chamber, and (b) the proximity of the left atrioventricular valve to the outlet chamber and hence the bulbouventricular foramenum. Some pulmonary venous blood in these patients appears to take a short cut through the common ventricular chamber to the aorta.

Interesting as these observations are, the results of stepwise regression analysis demonstrate that the major predictors of systemic arterial O2 saturation are pulmonary and systemic blood flow. Given these two factors, prediction of SA O2 saturation was not significantly improved by additional anatomic information. Thus, although in a few individuals the degree and direction of streaming may modify systemic arterial saturation to an important degree, in the group as a whole, the effect of streaming is slight by comparison with the effects of pulmonary and systemic blood flow. It was recently reported in a group of 39 catheterized patients with common ventricle and malposition (in whom the pulmonary artery was entered in 12) that the mean systemic arterial O2 saturation was 82.5% in l-malposition and 75% in d-malposition, whereas the mean pulmonary arterial O2 saturation was 75% in l-malposition and 85% in d-malposition.13 These figures at first sight suggest favorable streaming in l-malposition and unfavorable streaming in d-malposition. However, it was not made clear whether these means referred to 12 patients or to 39, what the simultaneous saturations were in individual patients, or what were the pulmonary and systemic flows. Thus we find these results difficult to interpret in the light of our own.

In the group with measured oxygen consumption 79% of the variation in SA O2 saturation could be predicted from Qp and Qs alone, whereas in the larger group including those with a predicted oxygen consumption these variables accounted for 13% less of the variation. This difference may well be wholly explained by the large random error of predicted oxygen consumption.18

In interpreting any statistical analysis one has to remember that correlation (or prediction) does not necessarily indicate causation. Interpretation of stepwise regression analysis carries the further hazard that a variable that is in fact causative may not prove to be predictive if either (a) its effect is masked by that of another closely correlated variable which is picked first by the computer or (b) it cannot be introduced in a suitably accurate form into the equation. Problem (a) can be, and was, largely overcome by inspecting correlations between independent variables and not introducing them into the same equation if they were closely correlated. Problem (b) can be overcome to a certain extent by examination of residuals, i.e., deviations of individuals from the predictive equation, as was done, but nevertheless is probably responsible for the failure to correlate pulmonary outflow gradient with systemic arterial saturation. It is intuitively obvious that the degree of pulmonary stenosis, by modifying Qp, affects SA O2 saturation. This is supported by the finding of a significantly lower SA O2 saturation and Qp in patients with pulmonary
stenosis than in those without pulmonary stenosis or severe pulmonary vascular disease. Pulmonary outflow gradient is presumably not only determined in common ventricle by the degree of pulmonary outflow obstruction, but also among other things, by the left atrial pressure, (particularly when pulmonary stenosis is severe) and by the ventricular systolic pressure (which is in turn dependent on the state of the systemic circulation). Therefore it has certain limitations as a measure of pulmonary outflow obstruction. It may be these limitations that prevented its being found predictive of SA O₂. By contrast, the role of Rp and Rp/Rs when pulmonary stenosis was absent was confirmed. The distribution of Rp against age was best fitted with a quadratic curve with a minimum during the second six months of life, indicating a qualitative similarity in natural history with ventricular septal defects. However, the behavior of any individual cannot be predicted from the limited data available.

Finally, the existence in our series of a patient with a systemic arterial oxygen saturation of 97.2% (P0₂ 86 mm Hg) re-emphasizes that the diagnosis of common ventricle will not be made unless it is thought of first, and then will only be made with certainty by angiocardiology. But after that, questions such as preoperative localization of specialized conducting tissue and chordal apparatus still need to be answered; we look to the day when this will be possible.

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