PULMONARY ATRESIA WITH BIVENTRICULAR AORTA

BY

SIMÓN MUÑOZ-ARMAS,* GUILLERMO ANSELMÍ,* MARÍA V. DE LA CRUZ,
AND IVAN MACHADO*

From Instituto Nacional de Cardiología de México

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Among the cyanotic congenital heart malformations, there is a group characterized anatomically by the presence of an atretic pulmonary artery and a biventricular aorta which acts as the only functioning vessel arising from the heart. A mixture of oxygenated blood from the left ventricle and unsaturated blood from the right ventricle is injected into the aorta, and the pulmonary vessels are filled from the aorta either through a patent ductus arteriosus, bronchial collaterals, or both. The main clinical features of patients bearing this type of heart malformation are deep cyanosis, single second heart sound, and decreased lung vascularity. A systemic pulmonary anastomosis is the most generally accepted surgical procedure of choice.

The term "pseudotruncus arteriosus" or "truncus aorticus" has been used by some authors (Brinton and Campbell, 1953; Cooley, Bahnson, and Hanlon, 1949; Humphreys, 1932; Kjellberg *et al.*, 1959; Kugel, 1931; Taussig, 1960) to designate those cases of atretic pulmonary artery and overriding aorta with pulmonary artery normally placed in front of the aorta, i.e. extreme forms of Fallot's tetralogy. In our opinion, this term should also include the much less frequent group of malformed hearts with atretic pulmonary artery and biventricular aorta but with transposition of the great arteries, since the basic hæmodynamics, in many ways similar to that of a true truncus, is essentially the same regardless of the spatial relation between the two great arteries.

The expression pulmonary atresia with biventricular aorta defines the basic anatomical and hæmodynamic features, and semantically separates these cases from those of pulmonary atresia with normal aortic root, which, anatomically, hæmodynamically, and clinically, constitute a totally different group.

It is the purpose of this paper to describe the anatomical aspects of 18 necropsy cases fulfilling the preceding definition, and to outline some anatomical types. A review of the published material on combined pulmonary atresia, biventricular aorta, and transposition of the great arteries is also presented.

MATERIAL AND METHODS

Eighteen hearts with pulmonary atresia and biventricular aorta were studied: eight from the Museum of the Instituto Nacional de Cardiología de México, representing 3 per cent of the 252 specimens; and 10 from the Museum of Pathology of the Hospital J. M. de los Ríos of Caracas, representing 4 per cent of the 250 specimens of congenital heart disease.

In each specimen the infundibulum of the right ventricle was studied, as well as the pulmonary valve, the calibre and length of the pulmonary artery trunk and its spatial relation with the ascending portion of the aorta, the degree of aortic overriding, the size of the branches of the pulmonary artery, and the thickness of the walls of both ventricles. Other associations were looked for and noted.

* Present address: Departamento de Cardiología, Hospital Universitario, Caracas, Venezuela.

FIG. 1.—Schematic representations of the two types of pulmonary atresia with biventricular aorta. (A) Without transposition of the great arteries (extreme Fallot's tetralogy). (B) With transposition of the great arteries. R.P.A., right pulmonary artery; L.P.A., left pulmonary artery; Ao.A, aortic arch; P.T., pulmonary trunk; Ao., aorta; T.C.S., trunco-conal septum; I.V.S., interventricular septum; R.V., right ventricle; L.V., left ventricle; C.S., crista supraventricularis; I, interventricular septal defect.

ANATOMICAL DESCRIPTION

For the purpose of anatomical description we will consider separately those cases of pulmonary atresia and overriding aorta without transposition of the great arteries—extreme Fallot's tetralogy—and those with pulmonary atresia, overriding aorta, and transposed great arteries.

Group I: Pulmonary atresia and biventricular aorta without transposition of the great arteries (extreme Fallot's tetralogy) (Fig. 1A). There were 15 specimens in this group (83%). The pulmonary artery of decreased calibre arose in every case to the right of the aorta in an anterior position, crossed it ventrally, and then turned back and leftward before dividing into the right and left pulmonary branches. The right anterior ventricular wall at the level of the infundibulum, immediately below the pulmonary artery, exhibits a prominent muscular bulge which gives the specimens a characteristic external aspect. They all showed marked hypertrophy of the right ventricle and an excessive length of the pulmonary artery trunk. The calibre of the pulmonary trunk and its branches was decreased in all cases. A ventricular septal defect was present in every case, behind the crista supraventricularis and immediately below the aorta, which overrode the interventricular septum in variable degree (33% overriding in 4 cases, 66% in 6 cases, and 100% in 5 cases).

In 7 cases the ductus arteriosus was patent; in 9 there was a patent foramen ovale, and in 2 there was an atrial septal defect in the region of the fossa ovalis.

Taking into account some important variations in the anatomy of the infundibulum of the right ventricle and of the pulmonary valve, 3 anatomical subgroups will be described, the recognition of which may prove useful for the diagnosis and helpful when surgical treatment is considered.

(A) *Atresia of the pulmonary valve and infundibular pulmonary stenosis* (Fig. 2A and B). There were 4 cases in this subgroup (26%). In all of them the crista supraventricularis was either of

FIG. 2.—(A) Pulmonary atresia and biventricular aorta without transposition of the great arteries (extreme Fallot's tetralogy). External aspect of the specimen. The pulmonary artery has a reduced calibre, arises in front of the aorta, and crosses it obliquely. (B) Internal aspect of the right ventricle of the specimen shown in A. Notice the stenosis of the infundibulum, the atresia of the pulmonary valve, and the overriding aorta. The probe shows the ventricular septal defect. (C) Pulmonary atresia and biventricular aorta with transposition of the great arteries. External aspect of the heart. The narrow pulmonary artery is posteriorly placed with respect to the aorta. (D) Internal aspect of the right ventricle of the specimen shown in C. The aorta arises from the infundibulum of the right ventricle in front of the crista supraventricularis. 1. Pulmonary artery; 2. aorta; 3. infundibulum of the right ventricle; C.S.-V., crista supraventricularis.

normal thickness or thicker than normal. The infundibulum of the right ventricle was very narrow, but an infundibular chamber was discernible. The pulmonary sigmoid valve cusps were totally fused causing an atresia of the pulmonary orifice.

(B) *Valvular pulmonary stenosis and aplasia of the infundibulum* (Fig. 3A). Only one case in our series (6%) had these characteristics. The infundibular cavity was absent because of the fusion of the parietal band of the crista supraventricularis with the anterior infundibular wall, forming a muscular mass with a thickness of about 20 mm. The pulmonary artery showed valvular stenosis but the arterial orifice was not atretic.

(C) *Atresia of the pulmonary valve and aplasia of the infundibulum* (Fig. 3B). This was the most numerous subgroup (10 specimens=66%). The crista supraventricularis was always muscular and it was fused with the anterior infundibular wall, thus producing the aplasia of the infundibulum. Both structures together formed an anteriorly placed muscular mass whose thickness ranged from

FIG. 3.—Pulmonary atresia and biventricular aorta without transposition of the great arteries (extreme Fallot's tetralogy). (A) Specimen illustrating the subgroup with infundibular aplasia and stenosis of the pulmonary valve. A section has been made in the anterior wall of the right ventricle in order to show the stenotic pulmonary valve arising directly from the muscular mass formed by the fusion of the crista supraventricularis and the anterior infundibular wall. (B) Specimen illustrating the group with aplasia of the infundibulum and atresia of the pulmonary valve. The right ventricle has been opened to show these anatomical features. (1) pulmonary artery; (2) aorta.

8 to 20 mm. with a mean of 13.3 mm. The pulmonary sigmoid valve cusps were fused, and the pulmonary artery actually arose from the muscular mass previously described.

Group II: Pulmonary atresia, biventricular aorta, and transposed great arteries (Fig. 2C and D). Three cases in our series belonged to this group (16%). It was characterized by the fact that the aorta arose in front of the crista supraventricularis and ran ventral and parallel to the trunk of the atretic pulmonary artery. The latter was placed behind and to the left, and it emerged as a blind sac from an atretic orifice. In two cases there was *laevocardia* with *situs inversus* and a persistent common atrio-ventricular canal of the complete form. In the third a ventricular septal defect behind the crista supraventricularis formed the pathway through which the right-sided aorta received the blood from the left ventricle.

DISCUSSION

In 1956 one of us (de la Cruz) proposed an ontogenic theory for the explanation of congenital truncoconal malformations. An account of the embryological processes involved in the production of the anatomical heart alterations we are dealing with was contained in that publication. A brief discussion of those developmental deviations will be included herein for a better understanding of the anatomical variations found in this series.

An extreme unequal partition of the truncus-conus, at the expense of the pulmonary artery, and malformation of the primordia of the pulmonary sigmoid valve cusps, are the embryological alterations common to all cases of pulmonary atresia with biventricular aorta. The unequal partition determines the small calibre of the pulmonary artery trunk and prevents the normal alignment of the truncoconal septum with the primitive interventricular septum, thus originating a ventricular septal defect, above which the aorta overrides the interventricular septum.

In the cases without transposition of the great arteries (extreme Fallot's tetralogy) there is a normal rotation of the spiral truncoconal septum (Fig. 1A). The truncoconal ridges arise with a normal clockwise rotation of 180° and, when they fuse with one another, they determine the formation of a spiral septum with the same type and degree of rotation. This makes the pulmonary artery branches (sixth aortic arch) which, at the cephalic end of the truncus, have a sinistro-dorsal position, communicate with the anatomically right ventricle which, in the caudal end of the conus,

has a dextro-ventral position. For the same reason the aortic arch (fourth aortic arch), which at the cephalic end of the truncus has a dextro-ventral situation, will communicate with the anatomically left ventricle, which in the caudal end of the conus has a sinistro-dorsal position (Fig. 1A). Thus, the pulmonary artery arises anteriorly and to the right crossing the aorta ventrally, thence to direct itself backwards and leftwards before dividing into right and left pulmonary artery branches.

Variations in the degree of unequal partition of the truncus-conus at the level of the conal region and of malformations of the primordia of the pulmonary sigmoid valve cusps bring about the existence of anatomical varieties.

In the cases with pulmonary valve atresia and infundibular pulmonary stenosis, the unequal partition of the truncus-conus is not severe enough at the level of the conal region to produce infundibular aplasia. The annulus of the pulmonary valve is very narrow and the primordia of the pulmonary sigmoid valve cusps present extreme alterations with complete fusion of the valve cusps and consequent pulmonary valvular atresia.

In the cases with infundibular aplasia and valvular pulmonary stenosis the degree of unequal partition of the truncus-conus is extreme at the level of the conal region and produces a complete fusion of the parietal band of the crista supraventricularis with the anterior infundibular wall: this produces aplasia of the infundibulum of the pulmonary artery. The valvular pulmonary ring is smaller than normal, leaving little space for the primordia of the pulmonary sigmoid valve cusps to grow; the latter fuse partially, originating valvular pulmonary stenosis.

In the cases with valvular pulmonary atresia and aplasia of the infundibulum the degree of unequal partition of the truncus-conus at the level of the conal region is of the same magnitude as in the previous group. The distortion of the primordia of the pulmonary sigmoid valve cusps is also extreme and results in valvular pulmonary atresia.

In the cases with transposed great arteries, besides the unequal partition of the truncus-conus and the alterations of the primordia of the pulmonary sigmoid valve cusps there is an abnormal development of the truncoconal septum. The truncoconal ridges lack their normal clockwise rotation of 180°; they emerge in a straight manner and when they fuse with one another a straight truncoconal septum is formed (Fig. 1B). This fact makes the pulmonary artery branches (sixth aortic arch), which at the cephalic end of the truncus have a sinistro-dorsal position, communicate with the anatomically left ventricle, which at the caudal end of the conus also has a sinistro-dorsal position. Similarly, the aortic arch (fourth aortic arch), which at the cephalic end of the truncus occupies a dextro-ventral position, will communicate with the anatomically right ventricle, which at the caudal end of the conus has a similar dextro-ventral situation (Fig. 1B). The aorta arises ventrally and to the right of the pulmonary artery, and both great arteries are parallel. The alterations of the primordia of the pulmonary sigmoid valve cusps are severe and lead to complete fusion and consequent atresia of the pulmonary valve.

Of our 18 cases of pulmonary atresia with biventricular aorta, 3 had transposed great arteries. This low incidence in comparison with the Fallot's tetralogy group is also found in the published reports. In reviewing the reports we found 14 cases of this group, and in 5 of them there was a complete description of the specimens (Fontana and Edwards, 1962; Kelsey, Gilmore, and Edwards, 1953; Kugel, 1931; Rossman, 1942; Young and Griswold, 1951): 1 had transposition of the great arteries with pulmonary atresia associated with a single ventricle and a single atrium (Rossman, 1942); in 3 the ventricular septal defect was described as occupying an anterior position immediately below the ventrally-placed aorta (Kelsey *et al.*, 1953; Kugel, 1931; Young and Griswold, 1951), and in a fifth case the exact situation of the ventricular septal defect was not defined (Fontana and Edwards, 1962). In 2 of our 3 cases there was an associated endocardial cushion defect and the ventricular septal defect was of the type commonly seen in this malformation, i.e. posteriorly placed, in the region of the atrio-ventricular canal. In our third case the ventricular septal defect was placed behind the crista supraventricularis, and the atretic pulmonary artery seemed to arise directly from the interventricular septum. This can be explained on the basis of a delayed disappearance of the conoventricular flange in this case, causing dextroposition of the truncus-conus,

TABLE I
PULMONARY ATRESIA WITH BIVENTRICULAR AORTA AND TRANSPOSITION OF THE GREAT ARTERIES

Author	Age	Associated malformations	Observations
Breschet (1826) ..	1 mth.	Dextrocardia, atrial septal defect, ventricular septal defect	Corrected transposition of the great arteries
Hickman (1869) ..	1½ mth.	Lævocardia, with situs viscerum inversus abdominalis atrial septal defect, ventricular septal defect	
Northrup (1889) ..	31 dy.	Dextrocardia, cor biloculare, patent ductus arteriosus	
Baumgarth (1902) ..	1 yr.	Dextrocardia, cor biloculare, patent ductus arteriosus	
McCrae (1905) ..	7 wk.	Lævocardia, with situs viscerum inversus abdominalis, atrial septal defect, patent ductus arteriosus	Corrected transposition of the great arteries
Beattie (1922) ..	Infant	Dextrocardia, atrio-ventricularis comunis complete variety, ventricular septal defect, patent ductus arteriosus	
Rösler (1930) ..	6 yr.	Dextrocardia, atrial septal defect, ventricular septal defect, patent ductus arteriosus	
Kugel (1931) ..	3 mth.	Ventricular septal defect, patent ductus arteriosus, agenesis of the right pulmonary artery	
Rossman (1942) ..	9½ mth.	Cor biloculare, atrio-ventricularis comunis, complete variety; patent ductus arteriosus	Corrected transposition of the great arteries
Miskall and Fraser (1946) ..	3½ dy.	Dextrocardia, cor biloculare, patent ductus arteriosus	
Mannheimer (1949) ..	7 yr.	Dextrocardia, cor biloculare, patent ductus arteriosus	
Young and Griswold (1951) ..	23 mth.	Lævocardia, with situs viscerum inversus abdominalis, patent foramen ovale, ventricular septal defect, patent ductus arteriosus	
Kelsey <i>et al.</i> (1953) ..	10 mth.	Dextro-rotation, patent foramen ovale, right-sided patent ductus arteriosus, single ventricle	Corrected transposition of the great arteries
Fontana and Edwards (1962) ..	6 mth.	Lævocardia with situs inversus, atrial septal defect, ventricular septal defect, patent ductus arteriosus	
Our Case (1) ..	1½ yr.	Ventricular septal defect, patent ductus arteriosus, patent foramen ovale	
Our Case (2) ..	7 mth.	Lævocardia with situs inversus, atrio-ventricularis comunis	
Our Case (3) ..	3 yr.	Lævocardia with situs inversus, atrio-ventricularis comunis	Corrected transposition of the great arteries

which, in turn, makes the ventricular septal defect appear behind the crista supraventricularis, and the atretic pulmonary artery arise in close relation with the interventricular septum.

It is interesting to point out that the great majority of the published cases had abnormal cardiac positions: 7 had dextrocardia, 4 had lævocardia, and 1 had dextro-rotation. In two of our cases there was lævocardia with situs inversus (see Table I).

Some other embryological facts are worth pointing out. In the first place it is important to emphasize that the branches of the pulmonary artery and the main trunk of this vessel have a different embryological origin. In fact, the branches originate from the sixth aortic arch and the trunk from the division of the truncus arteriosus. For this reason, even in cases in which the pulmonary artery is atretic and its trunk of reduced calibre, the right and left pulmonary branches may be of middle size or even near-normal size. In these a successful systemic-pulmonary anastomosis (Cooley *et al.*, 1949) can be performed even though the existence of pulmonary atresia could incline one to

expect a poor result. Of the 18 cases studied, 3 had practically normal-sized pulmonary branches; in 10 the pulmonary branches were smaller than normal, but even then larger than the pulmonary trunk; only 5 had very small pulmonary branches, too small to accept increased flow through a Blalock fistula or other type of systemic-pulmonary anastomosis.

In those cases with pulmonary atresia and biventricular aorta without transposition of the great arteries (extreme Fallot's tetralogy), there is no direct correlation between the degree of unequal partition of the truncus-conus and the degree of dextroposition (de la Cruz and da Rocha, 1956). This is expressed anatomically by the fact that, despite the extreme unequal partition of the truncus-conus with a very small pulmonary trunk in all cases, the overriding of the aorta was quite variable from one case to the other.

The recognition of different anatomical subgroups is important since the surgical possibilities are different for each one of them. In the group without transposed great arteries (extreme Fallot's tetralogy) the variations in the anatomy of the infundibulum of the pulmonary artery and the pulmonary sigmoid valve have important surgical implications. In those with a narrow infundibular chamber and valvular pulmonary atresia, a complete surgical repair may be indicated if other considerations do not contraindicate the procedure (Brinton and Campbell, 1953; Johns, Williams, and Blalock, 1953). Those cases with infundibular aplasia, with or without concomitant valvular atresia, are generally beyond the possibilities of a complete repair and they may only benefit from an anastomotic procedure. However, Lillehei (Lillehei *et al.*, 1955) operated on a patient with infundibular aplasia and atresia of the pulmonary artery, and he was able to do a complete correction with good results.

In those cases of atretic pulmonary artery, biventricular aorta, and transposed great arteries, the only surgical possibility is an anastomotic procedure of the Blalock type. This operation in cases with transposition of the great arteries with stenosis or atresia of the pulmonary artery, although with less favourable results than in Fallot's tetralogy, is followed by improvement in a good number of cases (Cleland *et al.*, 1957; Young and Griswold, 1951). Sometimes, even after angiocardiographic studies, the presence or absence of transposition of the great arteries cannot be established with certainty (Cleland *et al.*, 1957). In these, the diagnosis of pulmonary atresia with overriding aorta justifies the indication of the anastomotic operation regardless of the reciprocal position of the aorta and the pulmonary artery.

SUMMARY

The authors present an anatomical study of 18 specimens with pulmonary atresia and biventricular aorta. Two anatomical groups are distinguished according to the existence or absence of transposition of the great arteries. Three subgroups are outlined among the cases without transposition of the great arteries (extreme Fallot's tetralogy), determined by variations in the anatomy of the infundibulum of the pulmonary artery and the pulmonary sigmoid valves. The embryological deviations involved are discussed and the surgical possibilities of each anatomical group are analysed.

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REFERENCES

- Baumgarth, H. (1902). Cor biloculare mit Dexiocardie. Inaugural Dissertation, Halle.
 Beattie, W. W. (1922). Cardiac anomaly (bicameral heart) in situs inversus: mirror-picture transposition of ventricles, aorta, and pulmonary artery. Reversed insertion of aorta and pulmonary artery. Auricles not transposed. Superior and inferior venæ cavæ open in common persistent left ductus Cuvierii. Persistent ostium primum. Common auriculo-ventricular orifice. Ventricular septal defect. Atresia of reversed pulmonary artery. Patent right ductus arteriosus. Associated with: atresia of middle of œsophagus and communication of upper end of lower fragment with trachea. *Bull. int. Ass. med. Mus.*, 8, 219.
 Breschet, G. (1826). Mémoire sur l'ectopie de l'appareil de la circulation, et particulièrement sur celle du cœur. *Répert. gén. Anat. Physiol. path. Clin. chir.*, 2, 1.

- Brinton, W. D., and Campbell, M. (1953). Necropsies in some congenital diseases of the heart, mainly Fallot's tetralogy. *Brit. Heart J.*, **15**, 335.
- Cleland, W. P., Goodwin, J. F., Steiner, R. E., and Zoob, M. (1957). Transposition of the aorta and pulmonary artery with pulmonary stenosis. *Amer. Heart J.*, **54**, 10.
- Cooley, R. N., Bahnson, H. T., and Hanlon, C. R. (1949). Angiocardiography in congenital heart disease of cyanotic type with pulmonic stenosis or atresia. I. Observations on the tetralogy of Fallot and "pseudo-truncus arteriosus". *Radiology*, **52**, 329.
- de la Cruz, M. V., and da Rocha, P. J. (1956). An ontogenetic theory for the explanation of congenital malformations involving the truncus and conus. *Amer. Heart J.*, **51**, 782.
- Fontana, R. S., and Edwards, J. E. (1962). *Congenital Cardiac Disease: A Review of 357 Cases Studied Pathologically*, p. 192. Saunders, Philadelphia and London.
- Hickman, W. (1869). Malformation of heart; transposition of auricles and of aorta; absence of pulmonary artery; patent foramen ovale; communicating ventricles; with lateral transposition of principal viscera; cyanosis. *Trans. path. Soc. Lond.*, **20**, 88.
- Humphreys, E. M. (1932). Truncus arteriosus communis persistens. Criteria for identification of the common arterial trunk, with report of a case with 4 semilunar cusps. *Arch. Path.*, **14**, 671.
- Johns, T. N. P., Williams, G. R., and Blalock, A. (1953). The anatomy of pulmonary stenosis and atresia with comments on surgical therapy. *Surgery*, **33**, 161.
- Kelsey, J. R., Gilmore, C. E., and Edwards, J. E. (1953). Bilateral ductus arteriosus representing persistence of each sixth aortic arch. Report of a case in which there were associated isolated dextrocardia and ventricular septal defects. *Arch. Path.*, **55**, 154.
- Kjellberg, S. R., Mannheimer, E., Rudhe, U., and Jonsson, B. (1959). *Diagnosis of Congenital Heart Disease*, 2nd ed., p. 248. Year Book Publishers, Chicago.
- Kugel, M. A. (1931). A clinical and pathological study of two cases of truncus solitarius aorticus (pulmonary atresia). *Amer. Heart J.*, **7**, 262.
- Lillehei, C. W., Cohen, M., Warden, H. E., Read, R. C., Aust, J. B., DeWall, A., and Varco, R. L. (1955). Direct vision intracardiac surgical correction of the tetralogy of Fallot, pentalogy of Fallot and pulmonary atresia defects. Report of ten cases. *Ann. Surg.*, **142**, 418.
- Mannheimer, E. (1949). Morbus cæruleus: An analysis of 114 cases of congenital heart disease with cyanosis. *Bibl. cardiol. [Suppl. ad. cardiologia]*, **4**.
- McCrae, J. (1905). A case of congenital atresia of pulmonary artery with transposition of viscera: A second case of transposition. *J. Anat. (Lond.)*, **40**, 28.
- Miskall, E. W., and Fraser, J. A. (1946). Cor biloculare: report of a case. *Ohio St. med. J.*, **42**, 369.
- Northrup, W. P. (1889). Reptilian heart, or, rudimentary auricular septum, rudimentary ventricular septum, obliterated pulmonary orifice and trunk, absence of pulmonary veins, absence of mitral orifice, anomalous veins and arteries. *Proc. N.Y. path. Soc.*, 1888, p. 40.
- Rösler, H. (1930). Beiträge zur Lehre von den angeborenen Herzfehlern über die angeborene isolierte Rechtslage des Herzens. *Wien Arch. inn. Med.*, **19**, 505.
- Rossman, J. I. (1942). Cor biloculare with transposition of the great cardiac vessels and atresia of the pulmonary artery. Phylogenetic and ontogenetic interpretation. *Amer. J. clin. Path.*, **12**, 531.
- Taussig, H. B. (1960). *Congenital Malformations of the Heart*, 2nd ed., p. 275. [The Commonwealth Fund] Harvard University Press, Cambridge, Massachusetts.
- Young, M. D., and Griswold, H. E. (1951). Situs inversus of the abdominal viscera with levocardia. Report of eight cases submitted to the Blalock-Taussig operation. *Circulation*, **3**, 202.