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The Neonatal Intrapulmonary Arterial Pattern in Transposition of the Great Arteries*

A Microangiographic and Histological Study

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Das intrapulmonale, arterielle Gefäßmuster bei Neugeborenen mit Transposition der großen Arterien

Eine mikroangiographische und histologische Untersuchung

Zusammenfassung. An Lungenpräparaten von 10 Kindern mit isolierter Transposition der großen Arterien wurden mikroangiographische und histologische Untersuchungen (einschl. Serienschnittstudien) vorgenommen. Das Alter der Fälle betrug 5 Std bis 7 Wochen.

Bei Transposition zeigen die Lungenarterien insofern ein abweichendes Muster, als an den intralobulären Arterienästen entweder trichterförmige oder besonders stark gewundene Abschnitte zu beobachten sind. Abnorme, „abrupte" Pulmonalarterienäste von tintenfischähnlichem Aussehen kamen bei 3 Fällen vor.

Pulmobronchialarterien (Lumen 50—275 μ) wurden bei 3 Fällen beobachtet. Die meisten hatten abnorm ausgebildete Endarterien, die die Bronchuswand verließen und als Capillaren in den Alveolenwandungen endigten.

Bei allen Fällen war das Lungenparenchym reich an vergrößerten Bronchialarterien und vor allem an den sich in alveolare Capillaren verästelnden Bronchopulmonalarterien. Bei 3 Fällen wurden auch kleine supplementäre mediastinale Arterien beobachtet, die sich ähnlich wie die Bronchopulmonalarterien im Lungenparenchym verzweigten. An den Hauptästen der Bronchialarterien im Hilus war eine Vergrößerung des Lumens bis auf 1200 μ zu beobachten.

Die teilweise von Bronchopulmonalarterien versorgten Lobuli besaßen — in der Regel — capilläre Verbindungen mit pulmonalarteriell versorgten Nachbargebieten. Obliteration derartiger „kontralateraler" Pulmonalarterienäste lag in 2 Fällen vor, was darauf hinweist, daß die arterielle Versorgung des betreffenden Gebietes von den Bronchialarterien übernommen worden war. Bei 5 Fällen konnte eine kleinere Anzahl arterieller, bronchopulmonaler Anastomosen, vor allem solcher vom „end-to-end-Typus", beobachtet werden. Ihre funktionelle Bedeutung scheint jedoch ebenso gering zu sein wie die der Anastomosen der normalen neonatalen Lunge.

Summary. Microangiographic and histological studies including serial sectioning were carried out on lung specimens from 10 infants with isolated transposition of the great arteries. The ages of the subjects ranged from 5 hours to 7 weeks.

In transposition, the pulmonary arterial pattern deviates from the normal either by the presence of funnel-shaped portions of the intralobular arteries or by abnormal tortuosity of intralobular arteries. Abnormal, octopus-like abrupt branches of the pulmonary artery were demonstrated in 3 subjects.

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Pulmobronchial arteries (diameter range 50—275 μ) were observed in 3 subjects. Most of these arteries have abnormal terminal branches leaving the bronchial wall to end as alveolar capillaries.

In all subjects there is an increased systemic arterial supply of the lung tissue, particularly *via* numerous bronchopulmonary arteries ramifying as alveolar capillaries. In 3 subjects there were also small supplementary mediastinal arteries entering the pulmonary parenchyma to ramify like the bronchopulmonary arteries. The diameter of the main bronchial arteries in the hilus of the lung is increased up to 1,200 μ.

In lobules partly supplied by bronchopulmonary arteries, there are — as a rule — capillary communications with adjacent areas supplied by the pulmonary artery. Obliteration of such "contralateral" pulmonary-artery branches was demonstrated in 2 subjects indicating that the arterial supply of these areas has actually been taken over by the systemic arteries.

Arterial bronchopulmonary anastomoses were demonstrated in small numbers in 5 of the subjects. The most common type of anastomosis was end-to-end. As in the normal neonatal lung, these anastomoses probably have little functional significance.

This is the third part of a study on the pulmonary vasculature of the human fetus and infant. The findings in the two previous studies dealing with the normal intrapulmonary arterial pattern in the late fetal and neonatal period (ROBERTSON, 1967) and in infancy and early childhood (ROBERTSON, 1967) serve as a basis for the interpretation of the findings in the present report.

Transposition of the great arteries is incompatible with extrauterine life unless the anatomic conditions permit mixing of the systemic and pulmonary blood flows. In the absence of abnormalities of venous return and cardiac septal defects, these infants depend upon the shunting capacities of the foramen ovale and the ductus arteriosus, and they usually die within the first few postnatal weeks (TAUSSIG, 1960). A few children, however, have been reported to have survived to adolescence, even without a patent ductus arteriosus (ALEXANDER and WHITE, 1947; PUNG, 1955).

The present study was undertaken to investigate whether transposition of the great arteries, as an isolated cardiovascular malformation, is associated with any particular type of intrapulmonary arterial pattern which might offer an auxiliary shunt, or crossing-over, between the systemic and pulmonary circulations otherwise connected in parallell.

Preliminary results of this study have been reported earlier (ROBERTSON, 1964).

Previous Investigations

Previous structural investigations on the vasculature of the infant lung in transposition of the great arteries have mainly been concerned with the effects of pulmonary hypertension on the pulmonary arterial bed. Medial hypertrophy of muscular pulmonary arteries, intimal proliferation, thrombotic and plexiform lesions have been reported to occur from the age of 1 to 2 months, though particularly in cases with coexisting ventricular septal defect (NAEYE, 1963; WAGENVOORT et al., 1964; FERENCZ, 1964, 1966). Only a few reports have appeared on the bronchial arterial system in isolated transposition of the great arteries. Prominent dilatation of the bronchial arteries was recognized in neonatal and older infants (COCKLE, 1863; DORNING, 1890; CAMARRI and MARINI, 1965), but the intrapulmonary course of these abnormal bronchial arteries was not analyzed in detail.

Material and Methods

The material, which was collected from four hospitals from 1962 to 1965, consisted of lungs from 10 neonatal autopsy subjects with isolated transposition of the great arteries. The age of the subjects at the time of death ranged from 5 hours to 7 weeks.

In three subjects an artifical atrial septal defect had been established *ad modum* Hanlon-Blalock (BLALOCK and HANLON, 1950). One of these infants (A 87) died during operation, the other two survived the operation by one and two days. Postoperative complications (pericarditis, pleuritis, adrenal hemorrhage, cerebral edema) contributed to the fatal outcome in these two cases. In the remaining seven subjects, the cardiovascular malformation *per se* was considered the main cause of death.

The anatomy of the heart was essentially uniform throughout the series, except for the presence of an artificial atrial septal defect in three subjects. There was complete transposition of the aorta and the arteria pulmonalis without a ventricular septal defect. The hearts were enlarged with dilatation and hypertrophy principally affecting the right ventricle. Patency of both the ductus arteriosus and the foramen ovale was recorded at autopsy in eight subjects. In the remaining two subjects (ages: 10 days and 21 days), the foramen ovale was open but the ductus arteriosus anatomically closed (Table 1).

In no instance was there evidence of other than recent lesions of the pulmonary parenchyma — atelectasis, edema, hyaline membranes, intraalveolar hemorrhage, aspiration, focal pneumonia.

The pulmonary or bronchial arteries of the lungs were injected with 7.5 per cent aqueous suspension of fine barium sulphate (Micropaque®, Damancy & Co.). Tap water was used and the pH was 6.4. The ductus arteriosus — when not anatomically closed — was ligated before injection. The lungs were unexpanded and in atmospheric conditions during the injection procedure.

In five subjects the injection was made into the pulmonary arteries of each lung or into the pulmonary trunk. The injection pressure was continuously recorded and maintained around 80 mm Hg (60—100 mm Hg) in a non-pulsating flow. In four other subjects the injection was made into the thoracic aorta in order to fill the bronchial arteries of both lungs. The injection pressure in this group was kept around 100 mm Hg (80—120 mm Hg).

In the remaining subject the injection of the left lung was made into the pulmonary artery and that of the right lung into the thoracic aorta, with injection pressures of around 80 and 100 mm Hg, respectively. Injection time was at least 30 minutes in all subjects.

After injection, the lungs were fixed in 10 per cent neutral formalin for 4—7 days. Frontal slices of the lungs, 2—3 mm thick, were then radiographed and specimens were taken from all lobes of the lungs including the hilus. Regions with evidence of transmission of contrast from one arterial system to the other were particularly looked for and selected for microangiography. Evidence of such transmission was present in most of the specimens, rendering at least part of the bronchial arterial system visible in angiograms from pulmonary-artery injected specimens, and *vice versa*.

After paraffin-beeswax embedding, the selected specimens were cut into 1,500—2,000 μ thick blocks, which were stereomicroradiographed by a method previously described (LJUNGQVIST, 1963). An average of 11 pairs of microangiograms measuring about 3 × 5 cm were made from each pair of lungs. Areas of particular interest found in the microangiograms were cut out from the blocks and re-embedded in paraffin for histologic examination. The sections, 6—7 μ thick, were stained with Verhoff's or Weigert's elastic tissue stain and counterstained with van Gieson's stain. Serial sections were cut from all areas with evidence of arterial bronchopulmonary anastomoses or pulmobronchial arteries in the microangiograms and from other areas with an unusual appearance. An average of 9 blocks were serially sectioned from each pair of lungs.

Results

Pulmonary Arterial System

Basic Pattern. Microangiograms from the six pulmonary-artery injected specimens had two fairly distinct types of peripheral arterial pattern, here arbitrarily denoted as Type I and Type II.

Table 1. *Age, birth weight, cardiac surgery and main autopsy findings in 10 neonatal autopsy subjects with transposition of the great arteries*

Case no.	Age at death			Birth weight (g)	Cardiac surgery			Main autopsy findings			
	mth	d	hr		At age mth	d	Type	Heart weight (g)	Ductus arteriosus	Foramen ovale	Further observations
A 24			5	3490				23	patent	patent	Intraalveolar hemorrhage, focal pneumonia
A 62		1		3200				19	patent	patent	Moderate aspiration
A 63		1		3550				29	patent	patent	Moderate aspiration
A 87		4		3540		4	H-B	17	patent	(H-B)	Atelectasis, right lung
A 26		9		3200				28	patent	patent	—
A 120		10		2700				40	closed	patent	Enlarged orifices of bronchial arteries in the aorta. Atelectasis, right lung
A 59		11		3390				31	patent	patent	Pulm. atelectasis, focal pneumonia. Splenomegaly, 16 g (congestion)
A 121		21		4190		20	H-B	not recorded	closed	(H-B)	Pericarditis (staph. aureus). Post-op. pleuritis. Pulm. congestion + intraalveolar hemorrhage. Adrenal medullary hemorrhage
A 122	1	6		3860	1	4	H-B	53	narrow (patent to probe)	(H-B)	Cerebral edema. Hemorrhage in the tentorium cerebelli. Atelectasis + hyaline membranes, left lung. Mild intraalveolar hemorrhage
A 67	1	24		3260				52	narrow (patent to probe)	incompletely closed	Enlarged orifices of bronchial arteries in the aorta. Intraalveolar edema. Mild recent pneumonia

H-B = artificial atrial septal defect *at modum* Hanlon-Blalock.

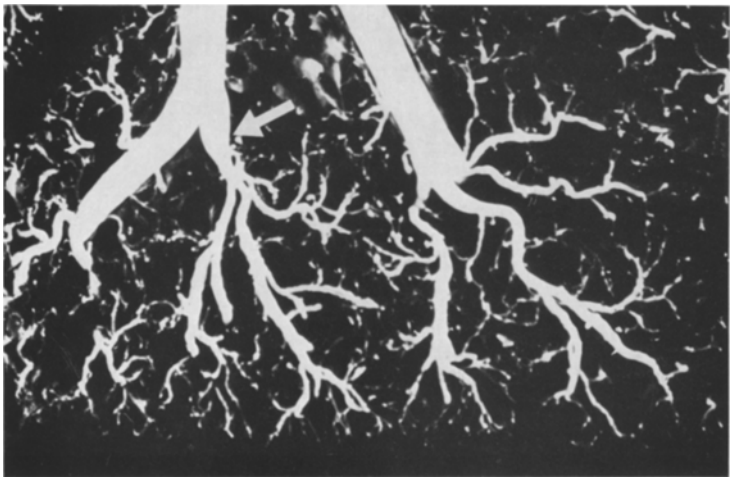


Fig. 1. Intralobular arterial pattern of Type I, with a rapid decrease in the diameter of the intralobular pulmonary arteries (*arrow*). Peripherally to these funnel-shaped portions, the arteries branches into bundles of ramifications of about equal size. Age 5 hours. Pulmonary-artery injected specimen. Microangiogram × 10

Type I, present in three subjects, is characterized by a rapid decrease in the diameter of the lobular pulmonary arteries at a level corresponding to the zone of transition of *mural structure* from *predominantly elastic to predominantly muscular*. Peripherally to the funnel-shaped narrowing the pulmonary arteries branch into bundles of small muscular arteries of about equal size, some of which even slightly wider than their parent artery (Figs. 1 and 3).

In *Type II*, present in the other three subjects, there is a more gradual decrease in the diameter of the pulmonary arteries from the hilus to the periphery of the lung. Deviation from the normal pattern is seen predominantly in the form of increased tortuosity of intralobular pulmonary arteries.

Table 2. *Basic pulmonary-artery pattern in 10 neonatal autopsy subjects with transposition of the great arteries*

Case no.	Age at death			Injection of	Type of intralobular pattern[a]	Endothelial, smooth muscle cushions in small muscular arteries
	mth	d	hr			
A 24			5	PA	I	+
A 62		1		PA	I	+
A 63		1		PA	I	+
A 87		4		BA	?	—
A 26		9		PA	II	—
A 120		10		BA	?	+
A 59		11		PA	II	—
A 121		21		BA	I (?)	+
A 122	1	6		PA, BA	II	+
A 67	1	24		BA	II (?)	—

[a] For definition, see text.

Abrupt muscular branches from elastic pulmonary arteries were frequent in all specimens and their frequency was unrelated to the Type I or Type II patterns. In three subjects (A59, A62, A63) the subdivisions of many of the abrupt branches form octopus-like bundles (Fig. 2); in one of these subjects (A59), the abnormal abrupt branches give rise to several pulmobronchial arteries. This feature will be further discussed below.

The secondary, incomplete filling of the pulmonary arteries obtained in most of the aorta-injected specimens did not allow a reliable microangiographic analysis of the fine intralobular arterial pattern (indicated by "?" in Table 2).

Prominent intimal cushions (ridges) of endothelial cells, occasionally also with a component of smooth muscle cells, were demonstrated histologically in six subjects (Table 2) and are particularly prominent in specimens with the Type I lobular arterial pattern. This feature appears segmentally in abrupt muscular branches immediately after their point of origin and in small intralobular muscular arteries, in Type I specimens usually just peripherally to the funnel-shaped portion of the parent vessel (Fig. 3).

A few pulmonary arterioles leave their lobuli to supply either septal tissue or the pleura or to anastomose end-to-end with septal or pleural branches of the bronchial arteries (see below). The diameter of these "penetrating" pulmonary arterioles, when distended with contrast, ranges up to 125 μ.

Pulmobronchial Arteries. Pulmobronchial arteries (i.e. intrapulmonary bronchial arteries originating from branches of the pulmonary artery) were demonstrated by serial sectioning in three subjects (30 per cent). These arteries send concurrent as well as recurrent branches along the bronchial wall. Their diameter at the point of origin ranges from 50 to 275 μ (Table 3). Some of the pulmobronchial arteries apparently substitute for, i.e. replace in the bronchial wall, bronchial arteries which then leave their usual position to ramify into the pulmonary parenchyma as "bronchopulmonary arteries" (see below).

The mural structure of the pulmobronchial arteries after they join the bronchial wall cannot be distinguished from that of ordinary bronchial arteries. They do not have the structure of "Sperr-arteries", i.e. their lumen is not narrowed by intimal bundles of smooth muscle cells.

Divergence from the normal pattern was observed in the following aspects:

a) Many of the pulmobronchial arteries deviated from the bronchial wall in their terminal course to ramify into alveolar capillaries (Table 3, Fig. 4). The diameter of these deviating branches ranges up to 100 μ.

b) In one subject (A59), the pulmobronchial arteries were particularly frequent and some of them arose as part of octopus-like ramifications of abrupt muscular branches from elastic pulmonary arteries (Fig. 2).

Bronchial Arterial System

Basic Pattern. In transposition of the great arteries the bronchial arteries generally follow the course of the bronchi, supplying the bronchial structures and lymph nodes. Pleural branches of the bronchial arteries are common, particularly at the mediastinal aspect of the lung and at the interlobar fissures.

Measured in the microangiograms, the inner diameter of the main bronchial arteries ranges from 375 to 1,200 μ (Table 4). In two subjects with particularly

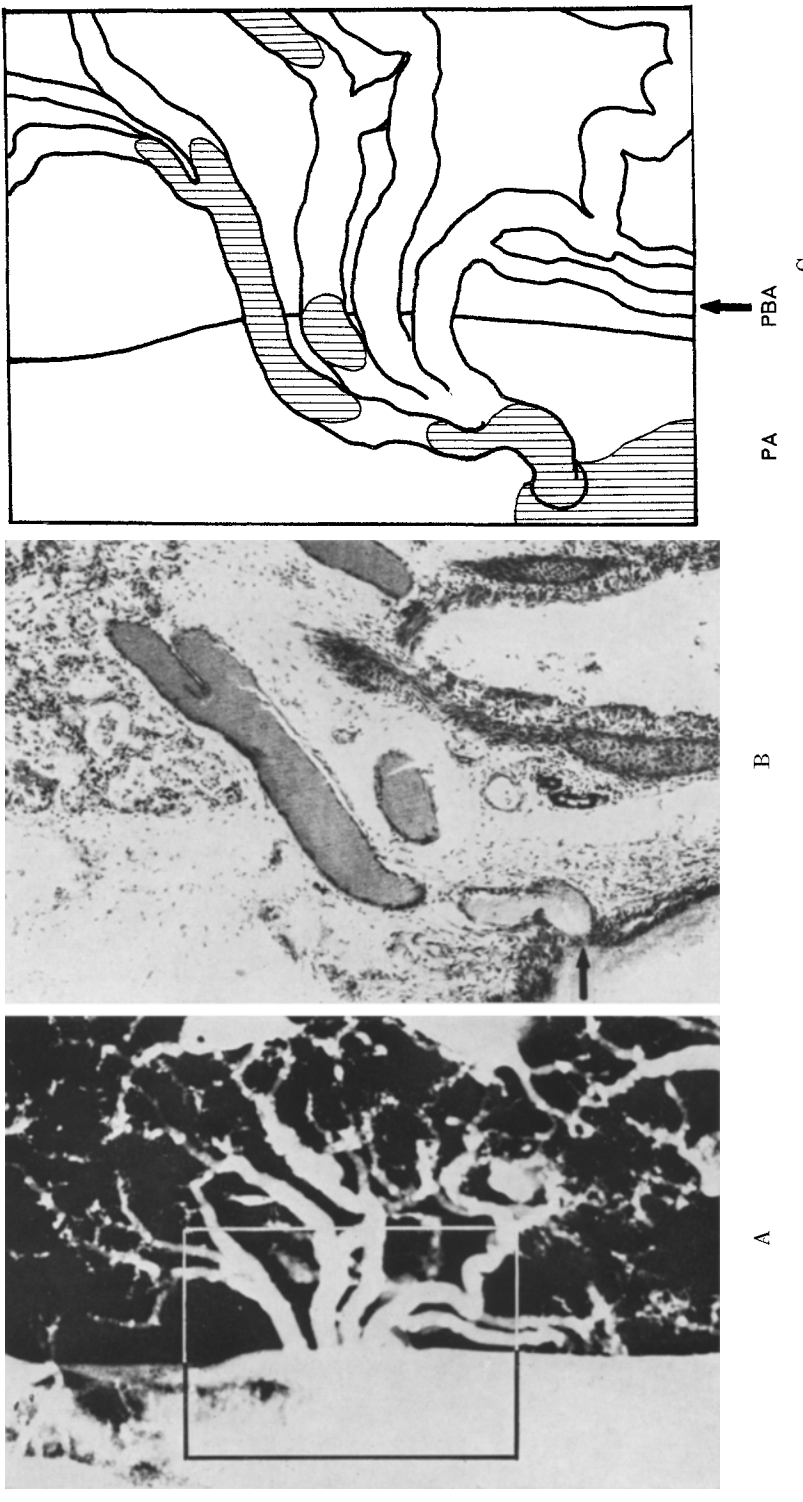
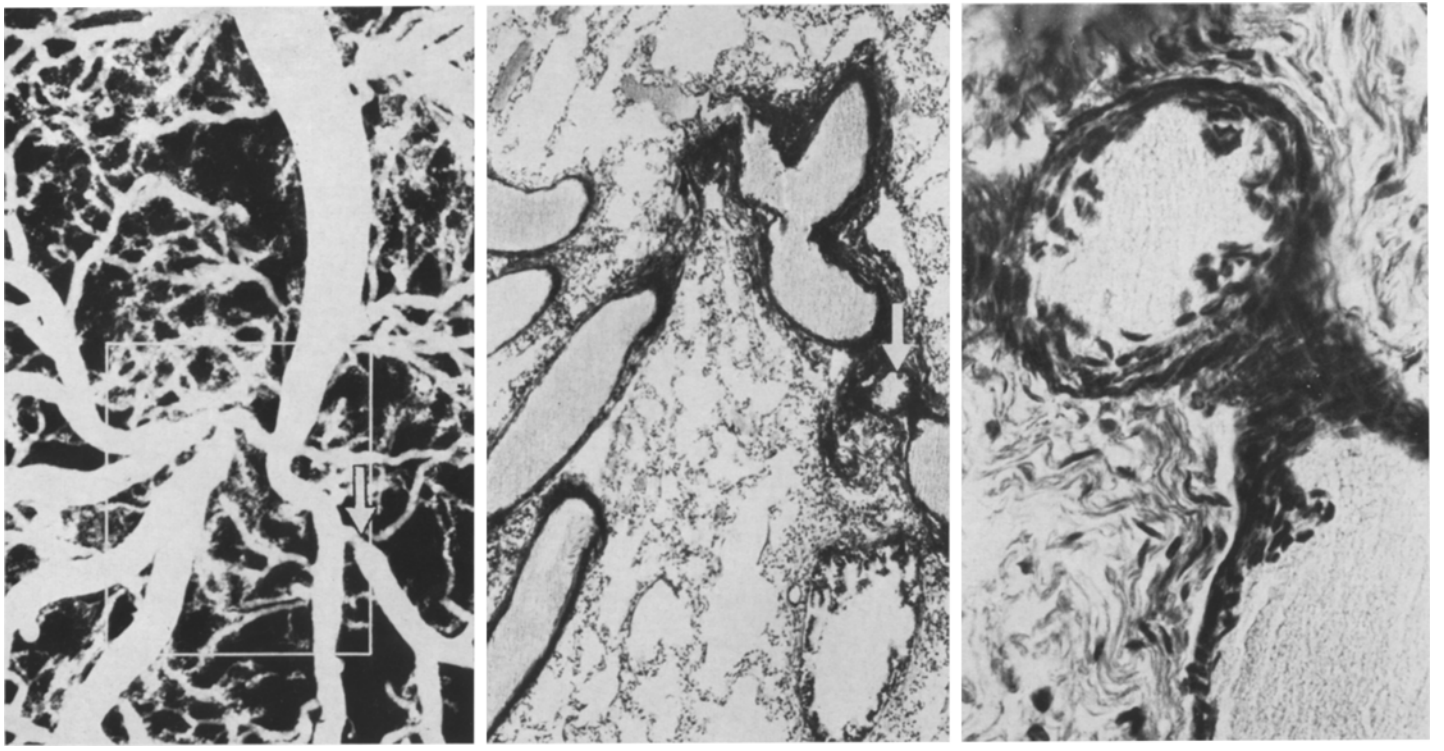


Fig. 2A—C. Octopus-like abrupt branch from the pulmonary artery (*PA*), combined with recurrent pulmobronchial artery (*PBA*, diameter 50 μ). Age 11 days. Pulmonary-artery injected specimen. A Microangiogram × 26. B Serial histologic section from framed area in the microangiogram, showing the point of origin of the abrupt branch (*arrow*). Weigert × 65. C Diagram of framed area in A, indicating the approximate level of B



A B C

Fig. 3 A—C. Intralobular arterial pattern of Type I with intimal cushions narrowing the lumen peripherally to the funnel-shaped portion of the pulmonary artery. Age 1 day. Pulmonary-artery injected specimen. Corresponding *arrows* in A and B. A Microangiogram × 26. B Serial histologic section from framed area in the microangiogram, demonstrating a bundle of branches of about equal size (*left*), and intimal cushions narrowing one other branch (*arrow*). Weigert × 65. C Higher magnification of the area indicated by *arrow* in B. Weigert × 420

Table 3. *Characterization of 15 pulmobronchial arteries demonstrated in pulmonary-artery injected lung specimens from three neonatal autopsy subjects with transposition of the great arteries*

Case no.	Age at death			Diameter (μ) of pulmobronchial artery	Site Lung lobe	Terminal branches to alveolar walls
	mth	d	hr			
A 62		1		150	LL	—
				50	RL	+
				225	RU	+
A 59		11		100	LL	+
				100	LU	—
				50	LU	—
				70	LU	—
				200	LU	+
				75	LL	—
				125	RM	+
				275	RL	+
				225	RL	+
				150	RL	+
A 122	1	6		75	LL	+
				75	LU	+

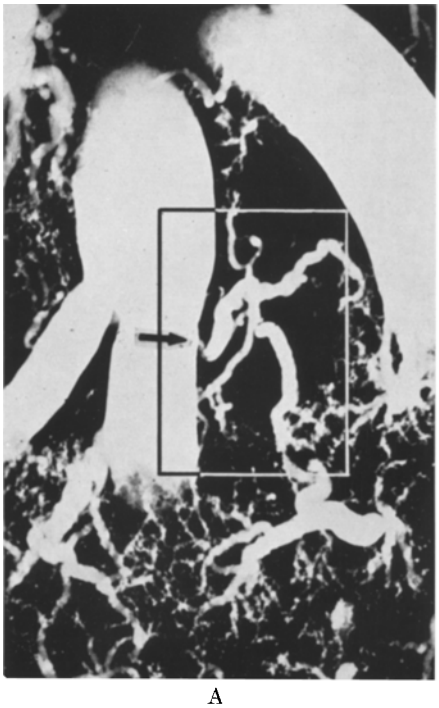
Table 4. *Diameter of the main bronchial arteries in the hilus of the lung in 10 neonatal autopsy subjects with transposition of the great arteries*

Case no.	Age at death			Injection of	Diameter (μ) of main bronchial arteries
	mth	d	hr		
A 24			5	PA	450
A 62		1		PA	450
A 63		1		PA	400
A 87		4		BA	550
A 26		9		PA	450
A 120		10		BA	750
A 59		11		PA	400
A 121		21		BA	625
A 122	1	6		PA, BA	375
A 67	1	24		BA	1200

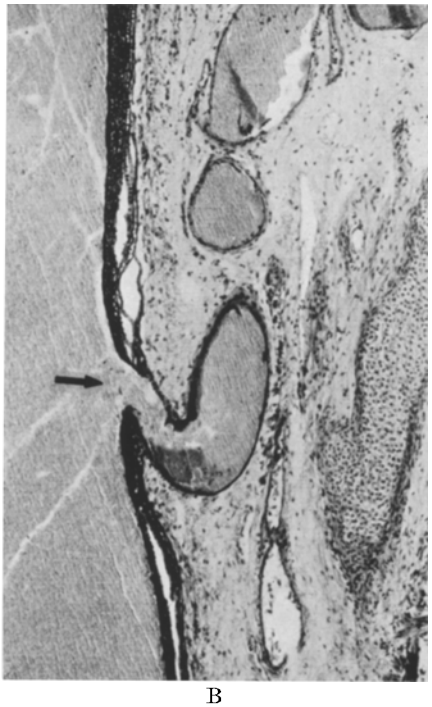
wide and tortuous bronchial arteries (A 67, A 120), enlargement of the aortic orifices of the bronchial arteries was recorded at autopsy.

In no instance did the bronchial arteries have the mural structure of "Sperr-arteries".

Bronchopulmonary Arteries. Bronchopulmonary arteries (i.e. branches from the bronchial arteries which enter the pulmonary parenchyma proper to ramify into capillaries of alveolar walls) are *numerous* in all subjects. These arteries are derived from intrapulmonary as well as pleural branches of the bronchial arteries. In some specimens the pleural bronchopulmonary arteries are prominent and easily recognizable to the naked eye at the mediastinal aspect and at the interlobar fissures



A



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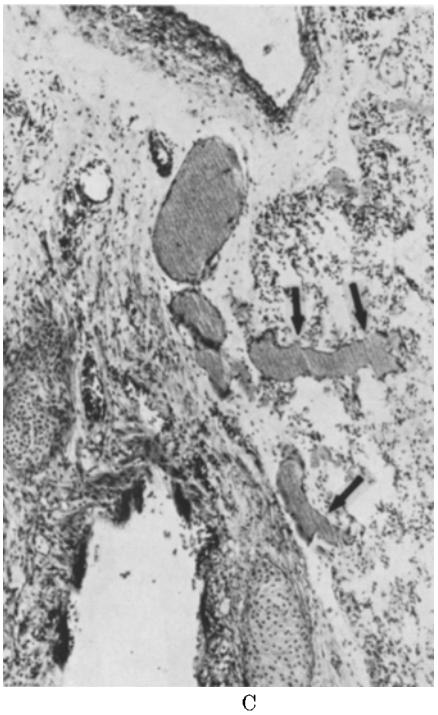

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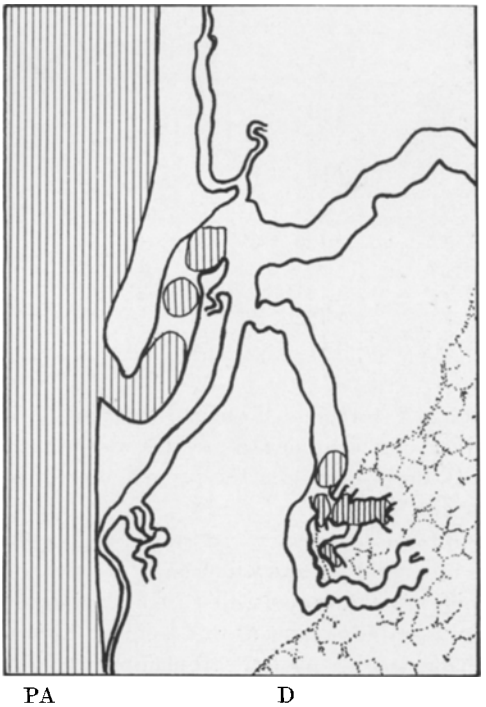


PA D

Fig. 4 A—D

Fig. 5. Enlarged and tortuous subpleural bronchopulmonary arteries visible to the naked eye at the interlobar fissure of the aorta-injected specimen. Age 10 days

of the injected lung (Fig. 5). Intrapulmonary bronchopulmonary arteries supply a large portion of the "medullary" (circumhilar) zone of the lung. In the aorta-injected specimes, a prominent capillary filling of the pulmonary parenchyma was generally obtained in this zone of the lung, whereas the alveolar capillaries of the periphery of the lung were poorly filled.

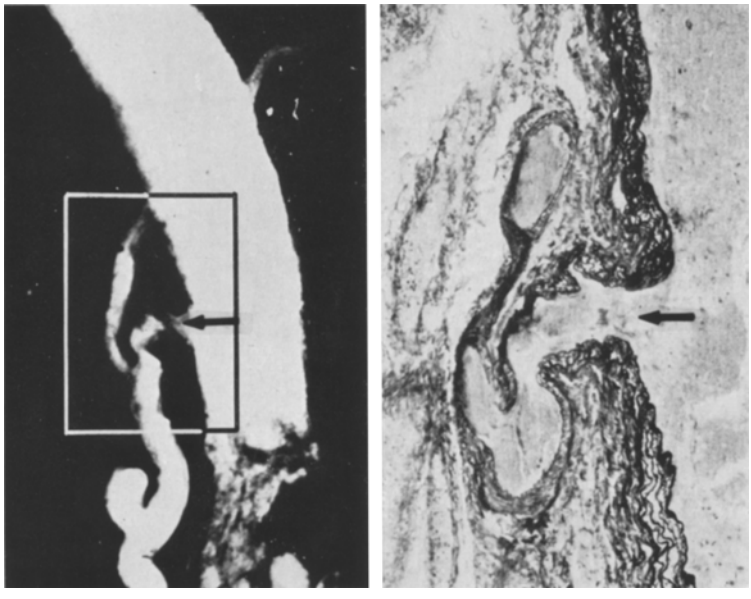
The inner diameter of the bronchopulmonary arteries, at their points of entrance into the pulmonary parenchyma, ranges up to 200 μ. The mural structure of these arteries after they enter the pulmonary parenchyma cannot be distinguished from that of ordinary pulmonary arteries of corresponding size. In no instance they had the structure of "Sperr-arteries".

In two instances a pulmobronchial artery originates not far distally from the point where a bronchopulmonary artery deviates from the bronchial wall, indicating that the former substitutes for the latter by taking over the arterial supply of the bronchial wall.

Supplementary Systemic-Artery Supply of the Lung

In three aorta-injected specimens with particularly wide bronchial arteries (A 67, A 120, A 121), the systemic-artery supply of the lung was further increased through the presence of multiple small mediastinal arteries, originating from branches of the thoracic aorta caudally to the bronchial arteries and contributing

Fig. 4 A—D. Pulmobronchial artery (diameter 150 μ) with abnormal terminal ramifications to the adjacent pulmonary parenchyma (C, *arrows*). The point of origin of the pulmobronchial artery is indicated by corresponding *arrows* in A and B. Age 11 days. Pulmonary-artery injected specimen. A Microangiogram × 10. B and C Selected serial histologic sections from framed area in A. Weigert × 62. D Diagram of framed area in A, showing the approximate levels of B and C. *PA* pulmonary artery, *PBA* pulmobronchial artery



A B

Fig. 6. Arterial bronchopulmonary anastomosis of side-to-side (*H*) type. The bronchial artery (*left*) is connected with the wider pulmonary artery (*right*) *via* a transverse vessel (diameter 100 μ). Corresponding *arrows* in A and B. In A, *upper*, the bronchial artery crosses the pulmonary artery. Aorta-injected specimen. A Microangiogram × 11. B Serial histologic section from framed area in A. Weigert × 29

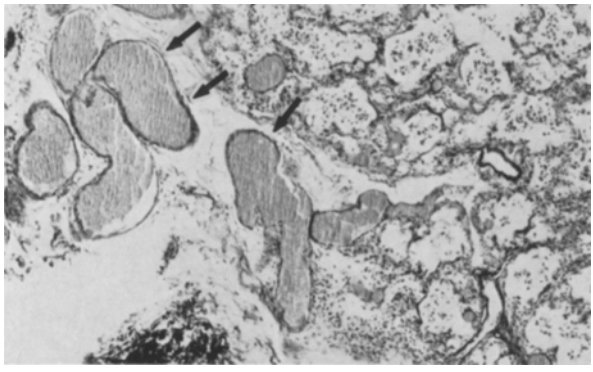
to the arterial supply of the neighboring dorsal portion of the lungs. In the injected specimens, these arteries were visible to the naked eye and entered the pulmonary parenchyma in the same way as the bronchopulmonary arteries.

The Relation between the Bronchial and Pulmonary Arterial Systems

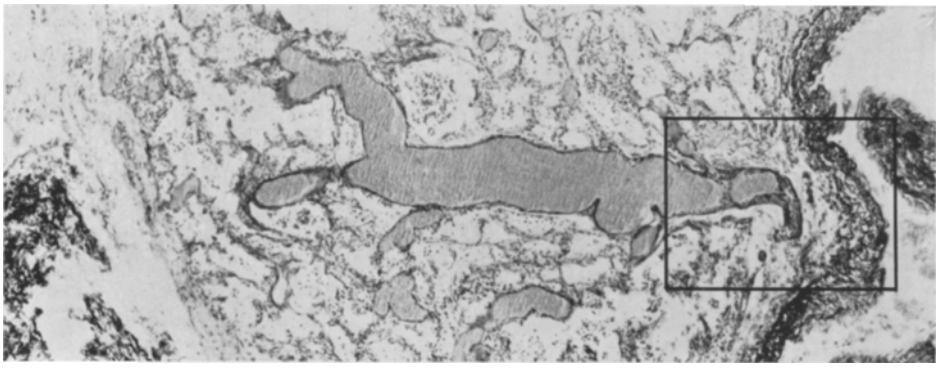
Arterial Bronchopulmonary Anastomoses. Arterial bronchopulmonary anastomoses were demonstrated by serial sectioning in five subjects (50 per cent). The anastomoses are few in number with not more than four in each subject. The most common type of anastomosis is end-to-end, in which a pleural or septal branch of the bronchial artery is connected with a pulmonary arteriole leaving (penetrating) a neighboring lobulus. Three anastomoses are of side-to-side type in which an intrapulmonary bronchial artery is connected with a pulmonary artery by a transverse vessel (Fig. 6A and B). No anastomoses of end-to-side type were encountered. The diameter of the anastomoses ranges from 50 to 125 μ (Table 5).

In the anastomoses there is a gradual change in the wall structure of the contributing vessels from bronchial-artery to pulmonary-artery type of mural elastic pattern. In the smaller anastomoses of end-to-end type there is an intermediate arteriolar or precapillary portion.

In one of the end-to-end anastomoses, present in one of the subjects with the Type I pattern, there was some stenosis of the lumen on the pulmonary-artery side through intimal cushions of the type described above. Otherwise, none of the



A



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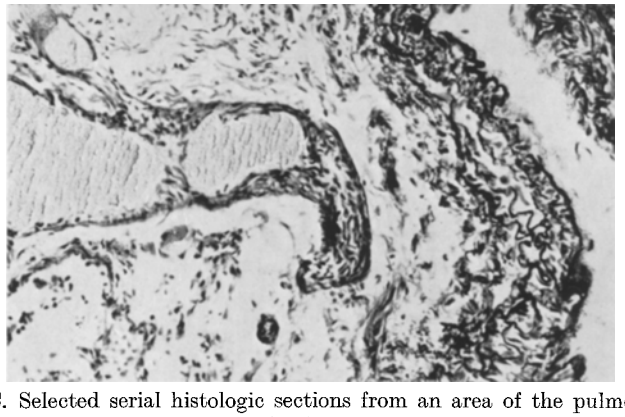

C

Fig. 7 A—C. Selected serial histologic sections from an area of the pulmonary parenchyma with a double arterial supply and with obliteration of the involved pulmonary artery. Age 10 days. Aorta-injected specimen. A A bronchopulmonary artery (*arrows*) runs from *upper left* to ramify into the pulmonary parenchyma (*right*). B Another level where the bronchopulmonary artery has entered the pulmonary parenchyma and anastomoses end-to-end with the "contralateral" pulmonary artery, running longitudinally across the field. The involved pulmonary artery is an abrupt muscular branch from an elastic pulmonary artery (B, C, *right*). This abrupt branch, however, is obliterated shortly after its point of origin by smooth intimal muscle cells. Serial sectioning demonstrated that the obliteration is complete. Weigert × 55. C Higher magnification of framed area in B. Weigert × 140

anastomoses displayed obliterative features, i.e. they did not have the mural structure of "Sperr-arteries".

Table 5. *Diameter and site of 10 arterial bronchopulmonary anastomoses, demonstrated in pulmonary-artery (PA) or bronchial-artery (BA) injected lung specimens from 5 neonatal autopsy subjects with transposition of the great arteries*

Case no.	Age at death			Injection of	Type of anastomosis, diameter (μ)			Site Lung lobe
	mth	d	hr		SS	ES	EE	
A 62		1		PA	—	—	100	LL
					—	—	125	RU
A 120		10		BA	—	—	100	LL
A 121		21		BA	100	—	—	LL
					50	—	—	LL
A 122	1	6		BA	—	—	50	RL
					—	—	50	RL
				PA	—	—	50	LL
					—	—	30	LL
A 67	1	24		PA	125	—	—	LL

EE = end-to-end; ES = end-to-side; SS = side-to-side.

The Relation Between the Bronchial and Pulmonary Arterial Systems in Areas of the Pulmonary Parenchyma with a Double Arterial Supply. In pulmonary lobules with a double arterial supply, i.e. lobules supplied by both systemic and pulmonary arteries, various forms of anatomic relation between the two arterial systems were encountered.

a) A common capillary network between the bronchopulmonary artery and adjacent branches of the pulmonary artery. This is the most frequent pattern, demonstrated in the microangiogram by the transition of contrast from one arterial system to the other.

b) Same as a), but with the additional presence of precapillary communications between the two arterial systems. These communications are included among the abovementioned arterial bronchopulmonary anastomoses of end-to-end type.

c) Hypoplasia or even obliteration — by intimal masses of endothelial and smooth muscle cells — of "contralateral" pulmonary-artery branches in areas further supplied by bronchopulmonary arteries (Fig. 7). This was demonstrated in two subjects (A120, age 10 days; A121, age 21 days).

Discussion

Previous studies of the structure of the pulmonary vasculature in transposition of the great arteries have revealed medial hyperplasia in small muscular arteries, thrombosis, fibrous intimal thickening and even plexiform lesions from the age of 1 to 2 months (NAEYE, 1963; FERENCZ, 1964). Tortuosity of the peripheral pulmonary arteries, which in the present study was a prominent feature of the Type II pattern, has been observed in post-mortem angiograms from cases of pulmonary hypertension secondary to other forms of congenital heart disease (SCHOENMACKER and VIETEN, 1954; DOYLE et al., 1957), and is apparently not specifically related to transposition of the great arteries.

The characteristic microangiographic appearance of the Type I pattern, with funnel-shaped segments of the intralobular pulmonary arteries, does not seem to have been recognized previously.

The intimal cushions of endothelial cells, bulging into the lumen of small intralobular or abrupt branches of the pulmonary artery in specimens with Type I pattern, are similar to the structures observed by v. HAYEK (1949, 1960) in the lungs of stillborn infants without cardiovascular malformations and referred to by him as "epithelioid" cells of "Sperr-arteries". On the basis of in-vitro experimental observations v. HAYEK claimed that the epithelioid cells swell under the influence of histamine to a sufficient degree to choke off blood flow through these vessels.

The functional significance of the two types of pulmonary arterial pattern cannot be assessed from the present study, but the rapid decrease in the diameter of the intralobular pulmonary arteries in the Type I pattern in combination with the apparently congenital intimal cushions of the same arteries suggests a higher vascular resistance than in the Type II pattern. This could be related with the observed difference in survival time between individuals with the Type I pattern (<1 day) and those with the Type II pattern (9 days — 5 weeks) provided that there is no evolution from one pattern to the other.

Octopus-like abrupt branches of the pulmonary artery, demonstrated in three subjects in the present series, have previously not been described in the human neonatal lung. The significance of these branches is unknown. Their association with pulmobronchial arteries could possibly be interpreted as a caricature of the normal pattern, in which many of the pulmobronchial arteries send small ramifications to adjacent alveolar walls, before they enter the bronchial wall (ROBERTSON, 1967).

The total incidence of *pulmobronchial arteries* in the present study (30 per cent) is close to the figure (24 per cent) obtained in a similar study on the late fetal and neonatal lung (ROBERTSON, 1967). The diameter range of the injected pulmobronchial arteries in the present series (50—275 μ), however, is considerably wider than in the normal neonatal lung (25—120 μ) (ROBERTSON, 1967; WAGENVOORT and WAGENVOORT, 1967). In a microangiographic study on the human lung in infancy and early childhood (ROBERTSON, 1967), pulmobronchial arteries with a diameter exceeding 250 μ were not demonstrated before the age of seven months.

The pulmobronchial arteries of the normal neonatal lung terminate — like the ordinary bronchial arteries — in the capillary plexus of the bronchial wall. In contrast to this, many of the pulmobronchial arteries in transposition of the great arteries deviate from the bronchial wall in their terminal course to end as alveolar capillaries. This is a challenge to the distinction between the pulmobronchial arteries and ordinary abrupt branches crossing the bronchial wall. An artery was classified as a pulmobronchial vessel, however, only if it passed through the cartilage of the bronchial wall.

The main bronchial arteries in the aorta-injected specimens of transposition hearts were wider (375—1,200 μ) than in the normal late fetal and neonatal lung (125—500 μ) or in normal infancy and early childhood (350—600 μ) (ROBERTSON, 1967).

Bronchopulmonary arteries, with a diameter ranging up to 125 μ, are consistently present in small numbers in the normal neonatal lung (ROBERTSON, 1967; WAGENVOORT and WAGENVOORT, 1967). It is evident from the present study that the bronchopulmonary arteries, in transposition of the great arteries, are considerably increased in number and that their neonatal diameter range is increased up to 200 μ. This feature, as well as the presence of supplementary mediastinal arteries running to the lung, is consistent with some previous reports on increased systemic arterial supply of the pulmonary parenchyma in transposition of the great arteries (COCKLE, 1863; ABBOTT, 1937; CAMPBELL and SUZMAN, 1951; LAPP, 1951; ASTLEY and PARSON, 1952; CUDKOWICZ and ARMSTRONG, 1952; EDWARDS, 1960; CAMARRI and MARINI, 1965; FERENCZ, 1966). The present study demonstrates that this abnormal systemic-artery supply is present at birth and that the systemic blood flow reaches the alveolar walls directly *via* bronchopulmonary arteries, and not *via* arterial bronchopulmonary anastomoses as has been suggested (COCKLE, 1863; CUDKOWICZ and ARMSTRONG, 1952). The pattern of a bronchopulmonary artery being replaced in the bronchial wall by a pulmobronchial artery, which was occasionally observed in this study, has also been recognized in the normal neonatal lung (ROBERTSON, 1967).

The incidence of *arterial bronchopulmonary anastomoses* in transposition (50 per cent) is greater than that of the normal neonatal lung (16 per cent) (ROBERTSON, 1967). In transposition of the great arteries, the most common type of anastomosis is end-to-end. This differs from the pattern of the normal infant lung, in which the most common type of anastomosis is side-to-side. The end-to-end type of anastomosis is close to the pattern usually present in areas supplied by bronchopulmonary arteries. In these areas there is, as a rule, a common capillary network between the bronchial and pulmonary arterial system (ROBERTSON, 1967). Since there is a gradual transition between the pattern of these capillary communications and the pattern of small anastomoses of end-to-end type, the increased number of such anastomoses is probably related to the increased number of bronchopulmonary arteries.

The total number of anastomoses, however, is small and since their diameter does not exceed 125 μ, they probably have little functional significance. A more important pathway for the systemic blood to the alveolar capillary bed is obviously offered by the numerous bronchopulmonary arteries. These vessels actually take over a considerable portion of the arterial supply of the lung tissue, which is further illustrated by the observation of obliterated "contralateral" pulmonary arteries in areas with a double arterial supply. This is contrary to the normal postnatal development in which the bronchopulmonary arteries obliterate and gradually disappear leaving the pulmonary arterial system as the only source of the arterial supply of the pulmonary parenchyma (ROBERTSON, 1967).

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