Etiology of Pulmonary Artery Dilatation and Hilar Dance in Atrial Septal Defect

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ABSTRACT

Atrial septal defect is associated with increased diameter and pulsation of the pulmonary arteries. The high pulmonary flow rate produces turbulence and a murmur. Intraluminal pressure is normal and vessel wall elastic properties may be altered as in poststenotic dilatation. Pulmonary artery angiograms in 6 normal patients, 9 with pulmonary stenosis and 10 with atrial septal defect were recorded on film, projected and traced. Arterial diameters at maximum systole and diastole were measured and, with intraluminal pressures, 2 points on the elastic diagram were calculated. These points lay on the initial linear portion of the graph so that a line through them to zero wall tension gave the resting radius (R₀). Each radius value (R) was normalized by dividing by the corresponding R₀. By plotting wall tension versus strain (R/R₀), a regression line was obtained for normals significantly different from that for poststenotic dilatation (P<0.01) and atrial septal defect (P<0.01). The slope of each line approximated vessel elastance at normal pressures: normals, 2.15 x 10⁴ dyne/cm; poststenotic dilatation, 1.00 x 10⁴ dyne/cm; and atrial septal defect, 0.89 x 10⁴ dyne/cm. Increased pulmonary artery distensibility extends more peripherally in atrial septal defect, thus producing “hilar dance.” A case of Marfan’s syndrome showed similar changes.

KEY WORDS
arterial wall elastic properties  turbulence
angiography  pulmonary stenosis  poststenotic dilatation
Marfan’s syndrome

Pulmonary artery dilatation and hilar dance are common findings in patients with atrial septal defects (1-8), but no satisfactory explanation has been given for either phenomenon. The theories suggested to explain the enlarged vessels include: congenital enlargement of the pulmonary artery, perhaps accentuated by the increased volume flow (2), increased volume flow from the right ventricle into the pulmonary artery (5, 6); and postnatal growth due to stimulation of wall receptors by the increased flow (9).

None of these suggestions accounts for all the changes seen. In general, only two factors will determine the diameter of a given vessel (if we neglect the small extravascular forces): (1) the intraluminal pressure acting perpendicularly to the wall in all directions, and (2) the elastic properties of the vessel wall which act circumferentially to resist distention of the vessel.

With an atrial septal defect, the diameter of the pulmonary artery is increased but the intraluminal pressure is usually normal (5, 6, 8, 10). This suggests that the elastic properties of the vessel wall might be altered so that the pulmonary artery is more distensible than normal. The situation would then be similar to another form of arterial dilatation, poststenotic dilatation. Here, altered elastic properties of the vessel wall are known to be responsible for the increased vessel diameter (11, 12).

To examine this analogy further, Roach previously showed that arteries will dilate distal to a stenosis only if a murmur (turbulence) is present (11, 13). Patients with atrial septal defect characteristically have
an ejection type of systolic murmur over the pulmonary artery (1-3, 7, 14). It arises from turbulence within the vessel, and Friedberg (15) attributes this murmur to the high flow rate, which produces a "relative stenosis" of the pulmonary valve.

Turbulence beyond a stenosis is localized to a relatively short distance. In atrial septal defect the nonlaminar flow probably extends well out into the larger branches of the pulmonary artery. This is suggested by the wide radiation of this murmur over the chest surface (16) and can be predicted from the mechanism of turbulence formation in any fluid. For a normal cardiac output and a pulmonary artery 2 cm in diameter, a common 2:1 shunt (pulmonary-systemic flow ratio) will produce a Reynolds' number exceeding the critical value of 2000 (17). Many patients with smaller shunts also have a murmur, which suggests that the Reynolds' number for a branching system of tubes may be less than that calculated for long straight tubes. This possibility has previously been suggested by Meisner and Rushmer (18) and Stehbens (19).

The similarity between the enlarged pulmonary vessels in atrial septal defect and the dilated vessel distal to a stenosis can be summarized as follows: Both lesions are associated with a normal intraluminal pressure, both show an increase in vessel diameter, and both have intravascular turbulence and a murmur. Since the vessel enlargement in poststenotic dilatation is due to altered elastic properties of the vessel wall (11), we must consider the possibility of similar changes being responsible for the dilatation seen in the pulmonary vessels with atrial septal defects.

We therefore compared the elastic properties of pulmonary arteries in atrial septal defect with normals and with dilated vessels distal to pulmonary valve stenosis. This comparison is facilitated by the normal low pressures within the pulmonary arteries, which means that the vessels operate on the initial (linear) portion of the tension-radius diagram. Greater changes to radius can be expected here between systole and diastole than are seen on the systemic side, where the pressure is higher and the artery less distensible (Fig. 1).

**Method**

The patients selected for this study had had routine cardiac catheterization and angiography. Radiopaque dye was injected into the outflow tract of the right ventricle or into the main pulmonary artery just distal to the pulmonary valve (20). The injections were recorded on 35-

mm film at a shutter speed of 50-64 frames/sec via either a 5-inch or a 9-inch image intensifier. The patient lay in a rotating cradle at least 60 cm above the x-ray source. With the input phosphor (receiver) located about 25 cm above the chest wall the magnification was about 1.5:1. The views taken were mainly in the posteroanterior position or the right anterior oblique, but the left anterior oblique was also used occasionally.

To avoid problems arising from alterations in the elastic properties of arteries with age (21, 22), no patient selected was older than 20 years (mean age 9.7). Patients were divided into three groups: (1) six patients with normal pulmonary arteries, mean age 11.7 years; (2) nine patients with moderate to severe pulmonary stenosis, mean age 8.7 years; and (3) 10 patients with atrial septal defects, mean age 7.3 years. The patients with pulmonary stenosis and atrial septal defect all had a murmur over the left sternal border at the second interspace.

In each case the film record of the dye injection was played back without knowledge of the intraluminal pressures. Two frames with good...
Normal Pulmonary Arteries

Two frames representing systole and diastole from the angiogram film of a male patient aged 15 with normal pulmonary arteries. Illustrated are the vessel outlines used for the diameter measurements and the pressure tracing from the main pulmonary artery (M.P.A.).

Diameters of the vessels at various points along their course were measured with a ruler (±0.5 mm). No measurements were made beyond the third branch of the left pulmonary artery or beyond the second bifurcation of the right pulmonary artery, since the dye was frequently diluted by these points and complete opacification of the vessels was not common. The catheter records were then consulted for the intraluminal pressure in systole and diastole recorded prior to the injection. Radiopaque dye injected into either the right or left heart raises systolic and diastolic pressures. This rise is about the same percent for systole and diastole and does not occur until over a minute after the injection (23, 24).

The pressures were obtained with a Statham model P23 series transducer (frequency response flat ±5% to 20 Hz) connected to the catheter lying within the pulmonary vessels and were recorded on either a Sanborn model 150 Polyviso, 4-channel recorder (frequency response flat 0 to 80 Hz, manufacturers specifications) or an Elec-...
Two frames representing systole and diastole from the angiogram film of a female patient age 7 with pulmonary valve stenosis and poststenotic dilatation. Dye injection was into the outflow tract, right ventricle. Illustrated are the vessel outlines used for the diameter measurements and the pressure tracing from the right pulmonary artery (R.P.A.).

tronics for Medicine DR8 (frequency response flat 0 to 30 Hz). In all cases, the pressures lay within the normal range; i.e., less than 30 mm Hg systolic (15).

Using the values thus obtained for systole (21.9 ± 3.8 mm Hg) and for diastole (8.1 ± 2.4 mm Hg) combined with the measured radii of the vessels (Fig. 5) two points on the elastic diagram were calculated from the law of Laplace ($T = P \times R$, where $T =$ wall tension, $P =$ intraluminal pressure, and $R =$ radius).

Although this method gives two numerical values for points on a tension-radius diagram for each artery, comparison of these values from one vessel to the next is complicated by several factors. First, the unstretched radius of any vessel decreases peripherally. Therefore, although an identically shaped elastic diagram will be present at any point along the course of a normal vessel, the actual numerical values for the wall tension and radius will differ. The same problem arises in attempting to compare these values from different patients. Finally, the degree of magnification of the vessels will vary slightly depending on the exact distance from x-ray source to patient and patient to receiver.

However, these pairs of points do represent a measure of the distensibility of the vessel over the normal pressure range. The slope of a line joining these two points would give a good approxima-
Two frames representing systole and diastole from the angiogram film of a male patient aged 15 with atrial septal defect. Dye injection in the outflow tract, right ventricle. Illustrated are the vessel outlines used for the diameter measurements and the pressure tracing from the right pulmonary artery (R.P.A.).
which has a rather low elastic modulus (27) and which is present in relatively small quantities in the large pulmonary vessels, cannot be assessed by our methods but does not show any histological changes in patients with atrial septal defect unless pulmonary hypertension has developed (28). Also, the initial slope of the elastic diagram appears to be very little altered by smooth muscle activity (29).

Results

For the six normal pulmonary arteries and branches a total of 15 separate systolic and diastolic diameters were measured and the corresponding wall tensions calculated. In addition a further ten similar measurements were made in seven patients with pulmonary valvular stenosis beyond the area of poststenotic dilatation. Each of these 25 pairs of values were plotted on a tension-radius graph and $R_0$ was obtained by projecting a line through the points to zero tension. Next, each value of radius was divided by the corresponding $R_0$. The stress-strain diagram could then be constructed by plotting wall tension ($T$) versus $R/R_0$ for all points (Fig. 6). The regression line calculated for these 50 points was $T = 21.5 \times 10^3 R/R_0 - 21.2 \times 10^3$ with a correlation coefficient of 0.82 and a standard error of the estimate of ± 0.87. The slope of this regression line, $2.15 \times 10^4$ dyne/cm, is the elastance for our normal pulmonary arteries, where elastance is Young's modulus times wall thickness (26).

The tension-strain diagram for the nine patients with pulmonary valve stenosis and poststenotic dilatation was obtained in the same manner (Fig. 6). The calculated regression line for the 48 points was $T = 10.0 \times 10^3 R/R_0 - 9.7 \times 10^3$ with a standard error of the estimate of ± 0.65 and correlation coefficient of 0.93. The elastance for pulmonary vessels with normal pressures and poststenotic dilatation was $1.00 \times 10^4$ dyne/cm, significantly different from normal ($P<0.01$). The standard method for comparing two regression lines was used (30).

For the third group of 10 patients with atrial septal defect, 25 systolic and diastolic radii were measured. The resulting plot of $T$ versus $R/R_0$ (Fig. 6) had the regression

![Plot of measured vessel radius at maximum systole (solid circles) and diastole (open circles) versus the corresponding systolic or diastolic pressures.](image-url)
Tension-strain diagrams. Tension is used rather than stress because wall thickness was not measured. The points representing wall tension versus normalized radius ($R/R_0$) give the regression line and standard error of the estimate shown. Elasticity for normal pulmonary arteries approximates the slope of this line, $2.15 \times 10^4$ dyne/cm; for pulmonary stenosis it is $1.00 \times 10^4$ dyne/cm; and for atrial septal defect, $0.89 \times 10^4$ dyne/cm. The tension-strain relation is given by the equation $T = 8.92 \times 10^3 \frac{R}{R_0} - 8.52 \times 10^3$ with a standard error of the estimate of $\pm 0.87$ and a correlation coefficient of 0.98. Therefore the elastance of the pulmonary vessels in atrial septal defect with normal intraluminal pressures was calculated to be $0.89 \times 10^4$ dyne/cm. This elastance is significantly different from normal ($P < 0.01$) but not different from that of poststenotic dilatation ($P > 0.1$).

One additional case, originally felt to fall into the "normal" group, since the angiogram and cardiac output were reported as normal, was of interest. The patient was studied because he had a low amplitude murmur over the pulmonary artery. All values of $T$ versus $R/R_0$ fell outside the normal range (Fig. 7) indicating increased distensibility of the vessels. Examination of this patient's old records showed a previous diagnosis of Marfan's syndrome. In 34 cases of this connective tissue disorder studied by Goyette and Palmer (31), a consistent deficiency of aortic elastin was noted.

There is no significant difference between atrial septal defect and pulmonary stenosis ($P > 0.1$).
fibers with replacement by loose collagenous tissue was noted. Involvement of the pulmonary arteries occurs (32, 33) but usually does not produce symptoms (34). Since elastin is responsible for the initial slope of the elastic diagram (26), the finding of abnormally distensible pulmonary vessels in our patient should not be entirely unexpected and further demonstrates the usefulness of this method.

Discussion

The slope of the elastic diagram for normal pulmonary vessels calculated by our method produced a value comparable to that available from Harris et al. (21). They measured pulmonary artery elastic properties by hanging weights on strips of the pulmonary vessels in vitro. Their youngest age group was 17 to 20 years with a mean age of 18.3 years (Fig. 1). From their results, we calculated a slope of $4.32 \times 10^4$ dyne/cm² for the section of their elastic diagram representing systole and diastole. This is about twice our calculated slope of $2.15 \times 10^4$ dyne/cm² for a normal group with a mean age of 11.7 years. However, age changes in the elastic properties of iliac arteries examined by Roach and Burton (22) showed that in five patients aged 0 to 10 years, the initial slope of the elastic diagram was less than half that drawn for three patients aged 11 to 20 years. Thus it is not surprising that the slope we calculated for our controls is about one-half that measured by Harris et al. (21) in their youngest age group.

The mechanical properties of the major pulmonary arteries in vivo were measured for 18 normal dogs by Patel et al. (35). For one dog of unstated age, they published an oscilloscopic plot of the pulmonary artery diameter (D) at various increasing pressures which showed the point $D_0$. Calculating values of $R/D_0$ from that graph we obtained values of wall tension versus strain similar to our results for the distensibility of normal human pulmonary vessels (Fig. 7). For their 18 dogs, an overall percent change in radius about the mean for a normal cardiac cycle was $\pm 7.8\%$. Our results showed an average normal radius change of $\pm 4.9\%$ about the mean. The radius changes of the pulmonary artery about the mean were considerably larger for poststenotic dilatation and atrial septal defect (± 12.1% and ± 12.9% respectively). Patel et al. also reported an average radius change of the right pulmonary artery in 30 human subjects of ± 8%. Their method was apparently similar to ours for measuring vessel radius on angiography, but the lesions present in those patients were not named.

Therefore the results obtained by our method for measuring the normal pulmonary artery distensibility do not appear to differ significantly from the results of other previously reported methods.

Our results show the pulmonary artery to be more distensible than normal distal to pulmonary valve stenosis, i.e., a lower slope for this portion of the elastic diagram and a slower rate of change of wall tension with increasing radius. This is as predicted by previous in vitro measurements of the elastic properties of vessels showing poststenotic dilatation (11-13). It also confirms that these changes are present in vivo. The increased distensibility of the vessel means that for any given pressure its radius will be greater than normal.

Finally, the pulmonary arteries in patients with atrial septal defect were more distensible than the controls, with a resulting increased diameter over their normal pressure range. Although the elastic properties are altered in the same way, the arterial dilatation with atrial septal defect extends more peripherally than that of poststenotic dilatation. This is because the turbulence in the pulmonary vessels with atrial septal defect is much more widespread, as suggested earlier.

These findings appear to rule out the previously proposed causes for the pulmonary artery enlargement. The increased volume being ejected into the pulmonary artery (5, 6) is seen only as an increased flow rate which cannot dilate low resistance vessels such as the large pulmonary arteries, but is indirectly responsible for the changes observed because it creates turbulence. Secondly, if the enlargement were congenital (3) the elastic properties of the wall should be normal. Also,
experimental production of atrial septal defects in dogs has shown the enlargement to be acquired (36). Finally, the suggestions made by Baltaxe and Amplatz (9) that the enlarged vessels are a growth phenomenon resulting from increased flow is also unlikely. Our results demonstrate that larger vessels maintain a normal, predictable distensibility. In addition, experimental work (38) demonstrated that the dilatation appears within weeks of the increased flow and is associated with the appearance of a murmur. The turnover rate of elastin is extremely slow (37) as is that of collagen (38) and it would seem likely that a much longer time, on the order of months, would be necessary for such growth to occur.

The only previous measurement of pulmonary artery elastic properties in atrial septal defect was done by Harris et al. (25). On the basis of in vitro tests of circumferential strips of pulmonary artery from two patients aged 34 and 45 years, they considered the elastic properties to be normal. Their method was to hang weights on the strip and to plot the predicted versus the observed percent extension. The histology in both cases showed an "adult pulmonary configuration" but also showed changes of "acquired pulmonary hypertension," which suggests that the media showed an increased thickness due to muscle hyperplasia (21, 28). They pointed out that "the extensibility is expressed in terms of unit cross-sectional area . . . the increased thickness of the wall . . . under these circumstances could by itself diminish its extensibility." Thus it is possible that they could miss the significant increase in pulmonary artery distensibility shown in our studies. Without pulmonary hypertension, our patients would all have a normal medial thickness, i.e., the "adult pulmonary configuration" which appears by age 2 (38).

Since we have shown that altered elastic properties of the pulmonary arteries can account for their increased diameter in atrial septal defects, one wonders if the abnormally great pulsation of the hilar vessels seen with fluoroscopy (2, 6, 8) can also be explained.

The average radius of the vessel is larger than normal and the slope of the elastic diagram is decreased over the range representing systole and diastole (Fig. 8). Therefore the radius change between these two normal pressures will be greater than normal (i.e., about twice normal for a reduction of the slope by one-half). This is seen on fluoroscopy as "hilar dance" or abnormally great pulsations of the hilar vessels. The localized nature of the dilatation with pulmonary valve stenosis does not create the same impression.

We would predict that hilar dance should decrease or even disappear under two conditions. First, as the pressure rises the artery would reach the elevated slope (collagen portion) of the elastic diagram occurring over 60 to 80 mm Hg (21, 39). This increased slope should be associated with decreased radius change between systole and diastole. In a study using electroneyography (40), patients with atrial septal defect showed pulsations of smaller amplitude if pulmonary hypertension was present. We found one patient aged 3 years with atrial septal defect and pulmonary hypertension of 100/40 mm Hg. Changes in radius between systole and diastole were small and the elastance was 18.5 X 10^4 dyne/cm, a value far greater than that seen at lower pressures.

![Figure 8](http://circres.ahajournals.org/)

**Figure 8**

Normal pulmonary trunk elastic diagram plotted from values given by Harris et al. (22). The decreased initial slope with pulmonary stenosis (P.S.) and atrial septal defect (A.S.D.) will produce greater radius changes ($\Delta R_n$) than normal ($\Delta R_n$) for normal intraluminal pressures. Extension of the dilatation to the hilum is interpreted as hilar dance.
Secondly, we might expect the hilar dance to decrease or even eventually disappear in older persons, since the tension-radius curve shifts to the left with age (21, 22). The initial slope gradually increases as more collagen is laid down and collagen crosslinking increases (22). Increased slope means decreased radius change between systole and diastole.

The one case of Marfan's syndrome warrants further discussion. This patient had a hereditary disorder of elastin which is characterized histologically by gradual elastin disintegration in the aorta (34) and pulmonary arteries (32). Since elastin contributes to the initial portion of the elastic diagram, its disappearance would also produce an increased distensibility of the vessel (decreased slope) and a greater radius change at normal intraluminal pressures. Thus, because of a murmur and turbulence, a patient with a congenital disorder of elastin can also present a picture similar to acquired dilatation.

References
PULMONARY ARTERY DILATATION


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