“Vanishing” pulmonary valve stenosis

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ABSTRACT

Objective: Both spontaneous resolution and progression of mild pulmonary valve stenosis (PS) have been reported. We reviewed characteristics of the pulmonary valve (PV) to determine factors that could influence resolution of mild PS.

Methods: Fifteen asymptomatic pediatric patients with spontaneous resolution of isolated mild PS were retrospectively reviewed.

Results: There was no correlation between the PV gradient, clinical presentation, age at diagnosis, or PV morphology. The PV annulus was small at initial presentation, which normalized at follow up. When corrected for the body surface area (z-score), the PV annulus was normal in all patients, including at initial evaluation.

Conclusions: Based on our observation, neither age at diagnosis, nor PV-morphology-influenced resolution of mild PS. The variable clinical presentation makes it difficult to categorize and observe mild PS by auscultation alone. The PV annulus z-score could be a useful adjunct to determine the course and serial observation of mild PS.

Keywords: Congenital heart disease, pulmonary stenosis, valvar disease

INTRODUCTION

Isolated pulmonary valve stenosis (PS) is a common congenital heart lesion with a perplexing natural history. The reported prevalence is 8–10% of all congenital heart lesions.[1] Although based on cardiac catheterization data, most natural history studies reveal no progression of mild PS.[2] Both spontaneous resolution of severe PS,[3] and progression of mild PS requiring intervention have been reported in infants.[4] Whereas some speculate lack of adequate growth of the PV orifice in PS,[5] we know linear growth of the PV orifice even in stenotic valves occurs with time.[6]

Recent natural history studies based on echocardiographic Doppler gradients are equally confusing. Again, lack of progression,[7] resolution,[8] as well as worsening of mild PS in neonates and young infants[9] has been reported. In general, there is lack of consensus on the natural history and the frequency of observation in patients with mild PS.[10,11] We reviewed the clinical and echocardiographic characteristics of the PV in patients with isolated mild PS that resolved spontaneously over time. An attempt was made to identify factors that could influence resolution of mild PS.

MATERIALS AND METHODS

The study was approved by the institutional review board of the University of Minnesota. Fifteen asymptomatic pediatric patients (<18 years at diagnosis) followed for isolated mild PS (PV gradient <40 mmHg), between November 1994 and August 2009 were selected. Two staff pediatric cardiologists at the University of Minnesota performed a retrospective review of charts, echocardiography studies, and echocardiography reports. Echocardiograms were reviewed on a Hewlett Packard Philips Sonos 5500 Ultrasound machine. All measurements were corrected for gender and body surface area and were represented as z-score values.[12] For echocardiogram images unavailable for review, information was extracted from patient charts and...
echocardiography reports. The patient inclusion criteria were clinical (at least a grade 2/6 systolic murmur or a murmur associated with a systolic click) or echocardiographic [peak PV gradient of ≥16 mm Hg or an abnormal PV (thickened or doming)] evidence of mild PS. On the last follow up visit, patients had no clinical or echocardiographic evidence to suggest PS.

RESULTS

Table 1 summarizes the clinical and echocardiographic findings of the 15 patients followed for mild PS.

At presentation: The age ranged from 2 days to 8 years with a mean of 11.8 ± 25.3 months, median 2.4 months. Mean PV gradient was 18 ± 8 mmHg (8–33 mmHg). The PV annulus measured 10 ± 3 mm (range 8–16 mm) with a mean z-score value of -0.17 ± 0.73 (-1.00 – 1.12). All patients had a murmur consistent with PS with a mean grade of 2/6 (range grade 1/6-3/6). Echocardiographic images for six patients were not available for review; however, reports were available, and were reviewed. Three patients had a systolic click at the pulmonary area associated with the murmur. Clinical data were not available for one patient. The patients were followed for a mean interval of 47.5 ± 38.8 months, median 36.5 months (range 4 months to 12 years).

At final evaluation: The patient age ranged from 4 months to 12 years with a mean of 59.3 ± 47.8 months, median 45.5 months. Mean PV gradient was 9 ± 4 mmHg (5–15 mmHg). The PV annulus measured 15 ± 3 mm (range 12–21 mm) with a mean z-score value of 0.08 ± 0.87 (-1.40 – 1.39). The mean grade for PS murmur was 1/6 (grade 0/6–2/6). Nine patients had improvement or complete resolution of the PS murmur, whereas, five patients had persistent PS murmur without any gradient by echocardiogram. There were insufficient data to comment on the clinical course in one patient. Only one of the original three patients continued to have a systolic click associated with the murmur.

DISCUSSION

The natural history of PS especially the milder forms remains contentious. With the emergence and improvements in echocardiography, it has become easier to make serial observations about changes in PS. Doppler echocardiography using a simplified Bernoulli’s equation is now regarded as a gold standard noninvasive method to determine severity of PS, since correlation between Doppler gradients and those obtained by catheterization has been established.

Even though the initial PV annulus diameter was smaller (10.2 ± 2.8 mm) compared to the final value (15.8 ± 2.5 mm), there was no difference when these

Table 1: Clinical and echocardiographic findings at initial and final evaluation of patients with mild pulmonary valve stenosis

| Patient | Initial age | Final age | Follow up | Initial PS murmur grade | Final PS murmur grade | Initial systolic click | Initial morphology | Initial PV morphology | Initial PV gradient (mmHg) | Final PV gradient (mmHg) | Initial PV annulus (mm) | Final PV annulus (mm) | Final PV annulus (z-score) |
|---------|-------------|-----------|-----------|-------------------------|---------------------|----------------------|--------------------|----------------------|--------------------------|--------------------------|-----------------------|-----------------------|--------------------------|
| 1       | 2 days      | 4 months  | 4 months  | 1                       | 1                   | No                   | Thick, doming      | Normal               | 12                       | 6                       | 8                     | NA                    | -0.6                    |
| 2       | 3 days      | 11 months | 10 months | 2                       | 2                   | Yes②                | Thick              | Normal               | 16                       | 14                      | 8                     | 12                    | 0.73                    | 0.11                   |
| 3       | 5 days      | 4 years   | 4 years   | 1                       | 1                   | Yes②                | Thick, doming      | Normal               | 21                       | 10                      | 8                     | 14                    | -0.6                   | 0.4                    |
| 4       | 1 week      | 5 years   | 5 years   | 3                       | 0                   | No                   | Thick, doming      | Normal               | 18                       | 10                      | NA                   | No echo               | 0.46                    | No echo                | 0.8                    |
| 5       | 3 weeks     | 19 months | 18 months | 2                       | 1                   | No                   | No echo            | Normal               | 10                       | 5                       | NA                   | No echo               | 15                     | 0.46                   |
| 6       | 1 month     | 12 years  | 12 years  | 3                       | 0                   | Yes                  | Thick, doming      | Normal               | 18                       | 9                       | 9                     | 21                    | -0.64                  | 0.14                   |
| 7       | 2 months    | 30 months | 2 years   | 3                       | 0                   | No                   | Thick              | Normal               | 23                       | 8                       | 10                    | 14                    | -0.9                   | -0.3                   |
| 8       | 2 months    | 3 years   | 3 years   | 2                       | 0                   | No                   | Thick              | Normal               | 14                       | 3                       | 9                     | 16                    | -1                    | -0.5                   |
| 9       | 2 months    | 3 years   | 3 years   | 3                       | 2                   | No                   | Thick, doming      | NA                   | 27†                      | 15                      | NA                   | NA                   | NA                    | NA                    |
| 10      | 4 months    | 3 years   | 3 years   | NA                      | 1                   | NA                   | Thick, doming      | Normal               | 9†                       | 9                       | NA                   | 14                    | NA                    | -0.16                  |
| 11      | 12 months   | 22 months | 11 months | 2                       | 2                   | No                   | Thick, doming      | Normal               | 33                       | 10                      | 11                    | 17                    | 1.12                   | 1.39                   |
| 12      | 12 months   | 9 years   | 8 years   | 3                       | 0                   | No                   | Doming†            | Normal               | 25†                      | NA                      | NA                   | NA                   | NA                    | NA                    |
| 13      | 15 months   | 9 years   | 8 years   | 3                       | 0                   | Yes②                | Doming†            | Normal               | 27†                      | 6                       | NA                   | 16                    | NA                    | -0.9                   |
| 14      | 24 months   | 4 years   | 2 years   | 2                       | 2                   | No                   | Thick              | Normal               | 10†                      | 9                       | 14                    | 19                    | 0.1                   | 1.3                    |
| 15      | 8 months    | 12 years  | 4 years   | 2                       | 0                   | No                   | Doming             | Normal               | 8                        | 5                       | 16                    | 16                    | -0.4                  | -1.4                   |

①: Identifies the patient with persistent systolic click at final evaluation.
values were corrected for the body surface area and expressed as z-scores (−0.17 ± 0.73 versus 0.08 ± 0.87, respectively). All the patients had normal (normal range −2.0 to ± 2.0) initial and final PV annulus diameter z-scores despite an abnormal appearing PV in many patients. It is possible that patients with progression of mild PS in previous reports had low PV annulus z-scores. Based on these findings, we believe that PV annulus diameter z-score values should be utilized in addition to PV gradients and physical exam to determine the management and follow up of patients with mild PS. Infants with mild PS and PV annulus z-scores of ≤-2.0 should be closely followed to monitor the adequate growth of the PV and resolution of PS.

Additionally in our study, we found no correlation between the PV gradient and the intensity of the systolic murmur, presence of a systolic click, or the PV morphology by trans-thoracic echocardiogram at initial evaluation. The fact that even a thickened, stenotic, and doming PV, with poststenotic dilatation of the main pulmonary artery demonstrated significant improvement on the last echocardiogram is very fascinating [Figure 1]. The intensity of the murmur decreased on follow up in most of our patients. Even though it is known that PS murmur intensity is proportional to the degree of valvular obstruction in moderate and severe PS, it would be difficult to categorize mild PS based on auscultation findings alone. The morphology of the PV in patients with mild PS has variable presentations unlike patients with moderate or severe PS. Whereas, some patients with significant PV thickening and doming generated minimal PV gradients, others with minimal abnormalities of PV morphology produced higher PV gradients and louder murmurs. This makes it difficult to predict the natural history of the spectrum of mild PS.

In general, no single factor determines the outcome of patients with mild PS. Although our study is limited by a small sample size, it raises valid questions and provides a basis for future investigations to determine the management and serial observation of patients with mild PS.

**CONCLUSION**

Based on our observation, there was no correlation between the severity of the PV gradient, age at diagnosis, clinical presentation, or the PV morphology in patients with mild PS. Because of this variable presentation, it is difficult to classify mild PS by physical exam alone. Though the initial PV annulus diameter was smaller than the normalized final values, when expressed as z-score even the initial values were normal for all the patients. We feel that the PV annulus diameter z-score could be a useful adjunct to the PV gradient and physical exam to help in determining the course and serial observation of mild PS.

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