ABSTRACT • OBJECTIVES: To describe the characteristics and outcome of children with an isolated silent patent ductus arteriosus (SPDA), with comparison to non-silent ducts.

PATIENTS AND METHODS: Between 1999 and 2004, all consecutive cases of isolated silent and non-silent patent ductus were recorded at the National Register of Pediatric and Congenital Heart Disease, Lebanese Society of Cardiology. Patients with a SPDA were followed clinically and by Doppler echocardiography while all non-SPDA were percutaneously or surgically closed.

RESULTS: Twenty-four cases of isolated SPDA and 50 cases of isolated non-SPDA ducts were recorded. Male sex was significantly predominant in the silent group (70%). First-cousin consanguinity rates were not different between both groups, with 20.4% for the silent group versus 22% for the non-silent group. Down’s syndrome was associated in three cases of SPDA. No cases of endocarditis were noted during a mean follow-up of 33.3 months. Four patients with a SPDA experienced spontaneous closure at the age of 25, 30, 36 and 58 months.

CONCLUSION: SPDA is a relatively benign disease. The risk of endocarditis cannot be totally ignored, but the systematic closure of the SPDA is not warranted. Larger series and longer follow-up are needed in order to draw conclusions. Spontaneous closure occurred in four patients with SPDA.

INTRODUCTION

A patent ductus arteriosus (PDA) is mostly audible as a continuous murmur in the first two left intercostal spaces at the left sternal border. A small patent arterial duct however, may not be audible on auscultation. In 1973, Abbott and Shively reported the first two cases of silent patent arterial ducts discovered on angiography [1]. The term “silent” patent arterial duct was then introduced in 1978 by McGrath et al., in preterm infants with respiratory distress and inaudible patent arterial duct [2]. This anomaly, not detected on auscultation, is discovered incidentally on routine echocardiography with color flow imaging done for various unrelated reasons [3]. By the end of the last century, with the advent of Doppler echocardiography, this entity called “the silent patent ductus arteriosus” (SPDA), was established among the medical community [3-4].

The management of these children is still controversial: Closure of the SPDA has been advocated to prevent the risk of infective endocarditis [5], but most authors recommend only observation [4, 6-7]. These opinions, however, are not based on clinical series or controlled trials. To date, there are only two published series about SPDA discovered in non premature, healthy infants or children [5, 8].

In this report, we describe the characteristics and outcome of 24 children with an isolated SPDA, along with comparison to non-silent patent ducts.
PATIENTS AND METHODS

Subjects

This series collected all consecutive patients with an isolated PDA, registered between 1999 and 2004 in the National Registry of Pediatric and Congenital Heart Disease, Lebanese Society of Cardiology. Inclusion criteria for an isolated patent ductus were:

1. Echocardiographic evidence of a patent arterial duct with typical color flow on cross sectional imaging and continuous systolic-diastolic high velocity flow into the pulmonary artery on pulsed Doppler.
2. Absence of any cardiac anomaly other than the patent arterial duct.

Patients were considered having a silent ductus if they met one additional criteria: the absence of any murmur or presence of soft or innocent murmur on auscultation.

Silent and non-silent PDA patients were compared for age, sex, size of the patent arterial duct, clinical symptoms, associated anomalies, consanguinity and outcome. Consanguinity was considered positive only if the mother and father were first order cousins. The size of the patent arterial duct was measured on ultrasonography in all 74 cases and on angiography only in patients with a non-silent patent arterial duct.

All silent patent arterial ducts were denied percutaneous or surgical closure. Prophylaxis against infective endocarditis was advised and regular systematic ultrasounds and clinical examinations were routinely performed every 6 months for follow-up. All non-silent patent arterial ducts were closed either percutaneously or surgically.

During the same period, consanguinity of all entries to the congenital heart disease registry were also analyzed for comparison.

Statistical analysis

All data are expressed as means and standard deviation, unless otherwise specified. Statistical comparisons between the two groups were done by unpaired t test or chi-square test. A p value less than 0.05 was considered statistically significant. We used SPSS software (version 9.0, SPSS Inc, Chicago, Illinois, USA).

RESULTS

Among 1480 entries for congenital heart disease, 74 babies had a patent arterial duct, 24 of which were silent. Hence, among all registered cardiac congenital anomalies, the incidence of isolated patent ducts was 5%, and the incidence of silent patent ducts was 1.6%.

Table I depicts the characteristics of all patients. Patients with a SPDA were younger but the difference was not statistically significant (p = 0.399). Surprisingly, there was a strong male predominance in patients with a SPDA (70% males) as opposed to patients with non-SPDA, with a significant statistical difference (p = 0.013).

The rate of consanguineous marriages in the whole group of patients with congenital heart disease (1480 patients), as expected, was higher than that of the normal population of Lebanon: 20.4% and 14.9% respectively [9]. Consanguinity in the silent duct group was similar to that of the 1480 patients of the registry. Consanguinity rate was slightly higher in the non-silent duct group (22%); the difference, however, was not statistically significant (p = 0.909).

Sizes of all silent ducts were smaller than those of non-silent ducts (inferior to 2 mm), with a mean diameter of one millimeter versus 4 millimeters respectively. Size correlated well with the clinical presentation: Patients with a silent duct presented for non cardiac problems, whereas those with a non-silent duct presented for typical cardiac symptoms.

Patients with a silent duct were brought to medical attention for various symptoms unrelated to the PDA:

- innocent murmur (50%)
- absence of murmur (50%): fever, nonspecific chest pain or palpitations, or bronchitis. The silent duct was also discovered during the overall assessment of children with Down’s syndrome or some other extracardiac anomaly.

No patient presented for signs of endocarditis during a mean follow-up of 33.3 months. Four patients (16.6%) with a SPDA experienced spontaneous closure at the age of 25, 30, 36 and 58 months. These unusual spontaneous closures prompted us to confirm patency retrospectively,
on at least two echocardiograms performed during the routine follow-up in all four cases. Figure 1 shows a spontaneous closure as confirmed by echocardiography.

**DISCUSSION**

Except for the premature infant, patent ducts accounts for 2.4% to 14% of all congenital heart diseases [10]. Whereas all non-silent ducts should be percutaneously or surgically closed, the management of a SPDA is still controversial [3, 8, 11-12]. Although it’s an asymptomatic duct with no hemodynamic or pulmonary consequences, concerns and questions persist about the risk of infective endocarditis. Without prophylaxis, the risk is around 0.45% per year and endocarditis was the most common cause of death of a patent arterial duct during the preantibiotic era [13]. For this reason, some authors recommend systematic transcatheter or surgical closure of a small patent arterial duct, whether it’s silent or audible [5]. Balzer et al. reported in 1993 the first case of endocarditis in a 19-year-old man with a clinically SPDA [14]. More cases of infected silent ducts followed that report [15-17], sometimes with unusual and life threatening complications such as concomitant pulmonary and aortic valve endocarditis in one patient, and aneurysm of the aortic isthmus in another patient. Moreover, an infected duct should be considered in the work-up of any child with unexplained prolonged fever and bacteremia: echocardiography is routinely performed in these children even in the absence of any previous cardiac history or murmur; it should be noted that even transesophageal echocardiography is not 100% sensitive, and the absence of vegetations does not exclude endarteritis [18].

Despite the risk of endocarditis however, most people simply recommend observation and antibiotics prophylaxis [6, 8, 19]. Cardiac symptoms are very unusual, and the rationale is to compare the small SPDA to a small residual leak following percutaneous coil closure of a patent arterial duct; such a residual shunt is conservatively managed and has been considered as a “benign techno-malady” which doesn’t need treatment nor prophylaxis [7]. Percutaneous coil closure of a small patent arterial duct carries a 2 to 14% risk of such a residual shunt [20-22].

The availability of a less invasive transcatheter method should not change the indications for intervention. Moreover, advocates of conservative management report more serious complications after coil closure attempts of silent ducts, not to mention the expenses [8, 23].

A SPDA duct is always an incidental finding. Unlike a large duct that may present with signs of heart failure, the silent duct presents for an unrelated reason that calls for a cardiac ultrasound [24-25]. On echocardiography, special attention should be given to the direction of the jet going into the pulmonary artery. Bennhagen and Benson showed that in most cases of silent ducts, the jet across duct may not reach the anterior wall of the main pulmonary artery, because of a narrower angle between the aorta and the duct [5]: the jet is thus diverted away from the anterior thoracic wall and doesn’t produce the typical audible murmur.

We witnessed four spontaneous closures of SPDA confirmed on echocardiography in children aged between 25 and 58 months. This unusual occurrence hasn’t been reported yet and shouldn’t be taken into account for the management of a silent patent duct.

**Figure 1.** Small PDA diagnosed at the age of one year: evidence of late systolic flow in the pulmonary artery (A); absence of flow during systole (B) and diastole (C) on the last Doppler echocardiography at the age of 3 years.
CONCLUSION

A debate still exists regarding the management of a SPDA. Despite the liberal use of antibiotics for infection today, the risk of endocarditis may still exist and should not be ignored. Although we didn't experience any complication related to conservative management of SPDA, larger series and longer follow-up without intervention are needed in order to conclude for long-term general safety and draw conclusions for the management of this relatively benign anomaly.

REFERENCES