

## Incidence of Congenital Heart Disease in Navarra (1989-1998)

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**Introduction and objectives.** Cardiac defects are the most common congenital malformation with an incidence of 5.2-12.5 per 1000 live births. The aim of this study was to describe the incidence and nature of congenital heart disease in the Spanish region of Navarra during a specified time period (1989-1998).

**Patients and method.** The study involved all children with congenital heart disease among the 47 783 born in the region in the specified time period.

**Results.** The incidence was 8.96 per thousand live births, with 90% having one of the 10 most common types of cardiac malformation. The accumulative percentage diagnosed was 25.3% in the first 24 hours of life, 45% in the first week, 65% in the first month, and 83.1% during the first year. Some 30.8% of cases of congenital heart disease required invasive treatment: 25.4% underwent surgery and 6.4% cardiac catheterization.

**Conclusions.** The incidence of congenital heart disease in Navarra falls within the range reported for developed countries. The level of care provided in this region is good, as demonstrated by existing diagnostic capabilities and treatment provision.

**Key words:** Congenital heart disease. Incidence. Extracardiac malformation. Syndrome. Cardiac surgery. Catheterization.

### Incidencia de las cardiopatías congénitas en Navarra (1989-1998)

**Introducción y objetivos.** Las cardiopatías congénitas son las malformaciones congénitas más frecuentes. Se detectan entre el 5,2 y el 12,5‰ de los recién nacidos vivos. El objetivo del presente trabajo es conocer la incidencia y la evolución de las cardiopatías congénitas en una región concreta de España (Navarra) y en un período determinado (1989-1998).

**Pacientes y método.** Se estudian los casos de cardiopatía congénita detectados entre los 47.783 niños nacidos en dicha comunidad durante el período indicado.

**Resultados.** Se detecta una incidencia de cardiopatías congénitas del 8,96‰ en recién nacidos vivos, el 90% de las cuales corresponde a las 10 malformaciones cardíacas más frecuentes. El porcentaje acumulado de diagnóstico es del 25,3% en las primeras 24 h de vida, del 45% en la primera semana, del 65% en el primer mes y del 83,1% durante el primer año de vida. El 30,8% de las cardiopatías congénitas requiere tratamiento invasivo: un 25,4% necesita cirugía y un 6,4% cateterismo terapéutico.

**Conclusiones.** La incidencia obtenida en Navarra está dentro del intervalo obtenido en los países desarrollados. Tanto la capacidad diagnóstica como el manejo evolutivo de las cardiopatías congénitas hacen de Navarra una comunidad con un nivel asistencial adecuado para dicha enfermedad.

**Palabras clave:** Cardiopatía congénita. Incidencia. Malformación extracardíaca. Síndrome. Cirugía cardíaca. Cateterismo.

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### INTRODUCTION

Cardiac defects are the most common congenital malformations. The incidence in industrialized countries ranges between 5.2 and 12.5 per thousand live births. The range is so wide because the estimation of the incidence depends on many factors, such as inclusion criteria, diagnostic means, size of the population, duration of follow-up, etc. In fact, in a review published in 2002, Hoffman et al<sup>1</sup> reported that the incidence of

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moderate and severe heart disease remains stable at about 6 per thousand, regardless of the place and time.

The objective of the present study was to determine the incidence and outcome of congenital heart disease in Navarra, a region in northern Spain with a population of somewhat more than half a million. An indirect assessment was made of the quality of the care offered to patients with congenital cardiac anomalies in this community in terms of diagnosis and treatment, taking into account the fact that heart surgery is not performed in the Community of Navarra and that children susceptible to invasive treatment must be transferred to another community.

## PATIENTS AND METHODS

A retrospective study was performed in the population of Navarra, with 523 563 inhabitants, over a 10-year period. An exhaustive search for cardiac malformations was carried out in all the public and private hospitals and primary care centers of the Community of Navarra, and in the hospitals offering cardiac surgery outside of Navarra to which patients requiring invasive treatment are referred. The authors investigated the presence of cardiac malformations among the 47 783 children born in Navarra from 1989 to 1998.

“Cases” considered suitable for inclusion in this study were all children born in Navarra between 1 January 1989 and 31 December 1998. The factors considered in relation to cardiac malformation were the need for invasive treatments, associated arrhythmias and, finally, a family history of congenital malformations or, more specifically, cardiac malformations.

The SPSS statistical software package (version 10.0) for Windows was used in the statistical study of the results. The differences were considered to lack statistical significance when the *P* value was more than .05, while a value less than .05 was considered to indicate a statistically significant difference and less than .01, a highly significant difference.

## RESULTS

### Incidence

The incidence of congenital heart disease in the study population was 8.96 per thousand live births. The different types of congenital heart disease recorded appear in Table 1. Ninety percent of those diagnosed corresponded to the 10 most common types of cardiac lesions. The relative frequency of each is shown in Figure 1.

### Sex

Among the population presenting heart disease, there was a slight predominance of girls (51.9%) over

boys (48.1%), although the difference was not statistically significant. All newborns were included, whether liveborn or stillborn, provided the gestational age was over 20 weeks. In some infants, the congenital heart disease was detected during the study period and, in others, later on. Data was collected up until 1 January 2003.

The definition of congenital heart disease was any anomaly in the heart or great vessels, among them, structural cardiac malformations, cardiac malposition not secondary to extracardiac malformations, congenital cardiomyopathy, and structural and vascular malposition not secondary to extracardiac anomalies. Atrial septal defects measuring less than 5 mm that closed prior to the age of 6 months, ductus arteriosus that closed during the first month of life (regardless of the gestational age), bicuspid aortic valve in the absence of aortic valve stenosis, mitral valve prolapse, cardiac malposition unaccompanied by structural heart disease and, finally, cardiac arrhythmias in the absence of other anomalies were excluded.

In the 428 children in whom congenital heart disease was diagnosed, the type of heart disease, the date and region of Navarra in which it was first detected, features of the delivery, and the diagnosis and course were studied. Among the children with patent ductus arteriosus, there was a highly significant predominance of girls ( $P=.003$ ), who also presented a significantly higher incidence of *ostium secundum* atrial septal defect

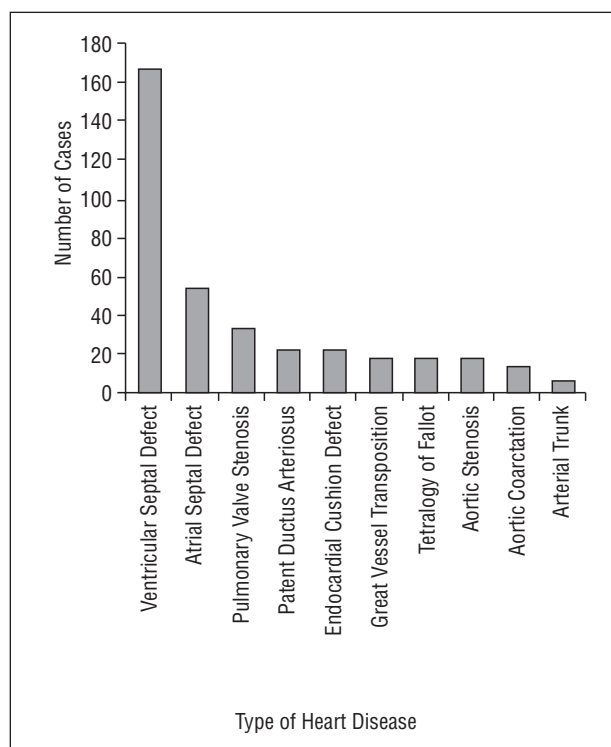


Figure 1. Most common forms of congenital heart disease.

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