analesdepediatría

www.analesdepediatria.org

ORIGINAL ARTICLE

Pulmonary hypoplasia: An analysis of cases over a 20-year period☆

Yanny Paola Delgado-Peña*, Alba Torrent-Vernetta, Gabriela Sacoto, Inés de Mir-Messa, Sandra Rovira-Amigo, Silvia Gartner, Antonio Moreno-Galdó, José Andrés Molino-Gahete, Felíx Castillo-Salinas

Hospital Universitario Vall dHebron, Barcelona, Spain

Received 31 August 2015; accepted 15 October 2015
Available online 19 June 2016

KEYWORDS
Pulmonary hypoplasia;
Pulmonary agenesis;
Congenital diaphragmatic hernia

Abstract
Introduction: Pulmonary hypoplasia is the most frequent congenital anomaly associated with perinatal mortality.
Material and methods: A retrospective and descriptive review was conducted on cases of patients diagnosed with pulmonary hypoplasia between 1995 and 2014 in a tertiary university hospital. An analysis was made of the prenatal imaging, clinical manifestations, post-natal diagnostic tests, treatment and management, long-term follow up, and survival data.
Results: A total of 60 cases were identified, all of them with prenatal imaging. Sixteen patients required foetal surgery. Congenital diaphragmatic hernia was the most frequent diagnosis. Main clinical presentation was respiratory distress with severe hypoxaemia and high requirements of mechanical ventilation. Mortality rate was 47% within first 60 days of life, and 75% for the first day of life. Pneumonia and recurrent bronchitis episodes were observed during follow-up. They had a lung function obstructive pattern, and their quality of life and exercise tolerance was good.
Conclusions: High neonatal mortality and significant long-term morbidity associated with pulmonary hypoplasia requires an early diagnosis and a specialised multidisciplinary team management.

☆ Please cite this article as: Delgado-Peña YP, Torrent-Vernetta A, Sacoto G, de Mir-Messa I, Rovira-Amigo S, Gartner S, et al. Hipoplasia pulmonar: análisis de la casuística durante 20 años. An Pediatr (Barc). 2016;85:70–76.

* Corresponding author.
E-mail address: yannypaola@yahoo.es (Y.P. Delgado-Peña).

PALABRAS CLAVE
Hipoplasia pulmonar;
Agenesia pulmonar;
Hernia diafragmática congénita

Hipoplasia pulmonar: análisis de la casuística durante 20 años

Resumen
Introducción: La hipoplasia pulmonar es la anomalía congénita más frecuentemente asociada a mortalidad perinatal.
Material y métodos: Se ha realizado un estudio descriptivo retrospectivo de los casos de hipoplasia pulmonar diagnosticados entre 1995 y 2014 en un hospital universitario de tercer nivel, analizando estudios prenatales, manifestaciones clínicas, pruebas diagnósticas, tratamiento, datos de seguimiento a largo plazo y supervivencia.
Resultados: Se identificaron 60 casos, todos con estudio prenatal. Dieciséis recibieron intervención quirúrgica intraútero. La hernia diafragmática congénita fue la entidad más frecuentemente asociada. La manifestación clínica más habitual fue distrés respiratorio neonatal, hipoxemia grave y necesidad de soporte con ventilación mecánica. Se halló un 47% de mortalidad antes de los 60 días de vida y del 75% en las primeras 24 h de vida. Durante el seguimiento de los supervivientes se detectaron episodios de neumonías y bronquitis recidivantes, función pulmonar con patrón obstructivo y aceptable calidad de vida y tolerancia al ejercicio.
Conclusiones: La elevada mortalidad neonatal y la importante morbilidad a largo plazo de la hipoplasia pulmonar requieren de un diagnóstico temprano y la intervención de un equipo multidisciplinar especializado.

Introduction

Pulmonary hypoplasia (PH) is a congenital anomaly characterised by impaired growth and development of the lung parenchyma, airways and vessels.[1–5] Its incidence in the general population is of nine to eleven cases per 10,000 live births,[2,3] although this must be an underestimate since infants with lesser degrees of PH survive the neonatal period.[2] The prevalence of PH reported in perinatal autopsy case series ranges between 7.8% and 26%,[1,3] and it is the anomaly most frequently associated with perinatal mortality.[1–6]

Pulmonary hypoplasia most frequently develops secondary to abnormalities in the thoracic cavity, foetal breathing movements, foetal lung liquid at positive pressure and volume of amniotic fluid.[1–7] Table 1 summarises and enumerates the underlying conditions most commonly associated to PH.[4,8,9] The clinical manifestations depend on the degree of lung involvement, ranging from severe bilateral forms to milder unilateral or lobar forms.[1,10] During the neonatal period, it can manifest with respiratory insufficiency, pulmonary hypertension or pulmonary haemorrhage. It characteristically presents with respiratory insufficiency of sudden onset that requires mechanical ventilation with high ventilatory pressures in the absence of atelectasis or obstruction.[2,11]

The diagnosis can be suspected prenatally based on measurements of lung volume by means of three-dimensional (3D) ultrasound and magnetic resonance imaging (MRI).[12,13] The diagnostic tests used after birth are thoracic computed tomography (CT) and lung scintigraphy, and the diagnosis is confirmed by pathologic examination.[1–4]

Different surgical approaches are used prenatally to attenuate the severity of PH in the foetus, such as drainage in cases of hydrothorax or foetoscopic tracheal occlusion (FETO) in severe cases of congenital diaphragmatic hernia.[14,15] After birth, the management of PH consists of supportive measures to guarantee adequate oxygenation during lung growth and development.

The established approach to the postnatal management of congenital diaphragmatic hernia is surgical repair followed by the use of invasive ventilatory support techniques with permissive hypercapnia combined with aggressive treatment for pulmonary hypertension.[10,15–18]

Recent studies have been pursuing the optimisation of the diagnostic methods, treatments and postnatal care used in patients with PH.[19] The aim of our study was to describe the aetiology, diagnostic methods and clinical outcomes of cases of PH diagnosed in a tertiary university hospital.

Materials and methods

We conducted a descriptive retrospective study of patients with PH diagnosed between 1995 and 2014 at the Hospital Universitario Vall d'Hebron de Barcelona that received care in the departments of obstetrics, neonatology, paediatric surgery and paediatric pulmonology.

We analysed the following variables: gestational age of the foetus at diagnosis, prenatal ultrasound abnormalities, prenatal surgery, gestational age at birth, age of onset of respiratory symptoms, clinical manifestations, aetiological diagnosis, diagnostic tests used, pathological findings, laterality of lung involvement, duration of required mechanical ventilation and oxygen supplementation, clinical respiratory outcomes (presence or absence of recurrent episodes of wheezing or infection), pulmonary function outcomes, quality of life and survival.