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ORIGINAL ARTICLE

Primary care follow-up of newborns with sickle cell disease detected in neonatal screening in the Community of Madrid☆

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Received 17 October 2013; accepted 7 April 2014
Available online 22 April 2015

KEYWORDS
Sickle cell anaemia;
Sickle cell disease;
Neonatal screening;
Care in primary care

Abstract
Objectives: The main aim of the study was to assess the effects of the recommended preventive programme in the population affected with sickle cell disease in primary care. The programme included, antibiotic prophylaxis, immunizations and health education, following the introduction of universal neonatal screening programme for sickle cell disease in the Community of Madrid.
Patients and methods: A cross-sectional observational study was performed with retrospective data collected from a cohort of newborns with sickle cell disease diagnosed by neonatal screening test in the Community of Madrid.
Results: From the data obtained from a sample of 20 patients, it was found that 95% had been diagnosed by the newborn screening test performed between 5 and 13 days of life. The mean age was 39 months when the study was conducted. During follow-up, from primary care paediatric clinic, it was observed that the compliance for antibiotic prophylaxis was 90%, and the coverage for the official vaccination schedule was 85%. Specific vaccine coverage as a risk population was highly variable (85% for pneumococcal 23 V, 50% for influenza, and 15% for hepatitis A). Health education only reached one in every four families.
Conclusions: Acceptable compliance with antibiotic prophylaxis was observed during the follow-up of patients with sickle cell disease in primary care, but a low coverage of routine immunisation, as well as specific immunisations. Coverage of health education was very low.
Improving these parameters would require greater coordination and involvement of primary care professionals so that these patients were followed up appropriately, and could be

☆ Please cite this article as: Rodríguez-Moldes B, Carbajo AJ, Sánchez B, Fernández M, Garí M, Fernández MC, et al. Seguimiento en Atención Primaria de los recién nacidos con enfermedad falciforme detectados en el cribado neonatal de la Comunidad de Madrid. An Pediatr (Barc). 2015;82:222–227.

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translated into a reduction of disease complications and an improvement in the quality of life of these patients.

PALABRAS CLAVE
Anaemia falciforme;
Drepanocitosis;
Cribado neonatal;
Cuidados en Atención Primaria

Seguimiento en Atención Primaria de los recién nacidos con enfermedad falciforme detectados en el cribado neonatal de la Comunidad de Madrid

Resumen
Objetivo: El objetivo principal del estudio fue evaluar desde Atención Primaria la repercusión del programa preventivo recomendado en la población afectada de enfermedad falciforme: profilaxis antibiótica, vacunaciones y educación sanitaria, tras la implantación del programa de cribado neonatal universal de anaemia falciforme en la Comunidad de Madrid.
Pacientes y métodos: Se realizó un estudio transversal observacional con recogida de información retrospectiva de una cohorte de niños con enfermedad falciforme diagnosticados por la prueba de cribado neonatal en la Comunidad de Madrid.
Resultados: Se obtuvieron datos de una muestra de 20 pacientes. El 95% de ellos fueron diagnosticados por la prueba de cribado neonatal realizado entre los 5 y 13 días de vida. La edad media de los pacientes cuando se realizó el estudio fue de 39 meses. En el seguimiento de estos pacientes desde Atención Primaria, se objetivó un cumplimiento de la profilaxis antibiótica del 90% y una cobertura del calendario vacunal oficial del 85%. La cobertura de vacunas específicas: 85% neumococo 23 V, 50% gripe y 15% hepatitis A. El reconocimiento de haber recibido educación sanitaria solo alcanzó a una de cada 4 familias.
Conclusiones: En el seguimiento de los pacientes con anemia falciforme en Atención Primaria, se objetivó un aceptable cumplimiento de la profilaxis antibiótica, pero unas bajas coberturas tanto de las inmunizaciones sistemáticas como de las inmunizaciones específicas. La cobertura de educación sanitaria fue muy baja.
Para mejorar estos parámetros, sería necesaria una mayor coordinación e implicación de los profesionales de Atención Primaria con el fin de que estos pacientes tengan un seguimiento adecuado y que ello se traduzca en una disminución de las complicaciones de esta enfermedad y una mejoría en la calidad de vida.

Introduction

Sickle cell disease (SCD) comprises a group of chronic inherited anaemias characterised by haemolysis and intermittent episodes of vascular occlusion leading to high morbidity and with a highly variable clinical expression.[1]

Early diagnosis allows for the combined followup by hospital paediatric haematology units and primary care (PC) from the first months of life. The various professionals involved are responsible for providing information about the disease and health education, initiating antibiotic prophylaxis, and ensuring compliance with the official routine and specific immunisation schedules.[2–5]

The aim of these measures is to improve quality of life significantly, reducing the number of complications and increasing the life expectancy of affected patients, as mortality due to SCD in the early years of life can reach up to 10% in the more developed countries.[4]

The most common presentation of SCD is the HbSS homozygous form, which affects approximately 75% of these patients. It manifests with severe haemolytic anaemia, vaso-occlusive bone pain crises, susceptibility to severe infections and severe end-organ damage, especially in the first 3 years of life. Other, less prevalent presentations are HbSC (25%), sickle thalassaemia (less than 1%) and sickle cell—other haemoglobinopathies, with a highly variable clinical expression, but usually milder.[5,6]

Clinicians in Spain are facing the challenge of treating SCD more often due to the increase in immigration, especially from African countries, Central and South America, and Asia, that has occurred in every autonomous community.[5,6]

In May of 2003, the autonomous community of Madrid (ACM) added a universal neonatal screening programme for the early detection of haemoglobinopathies to the already established screening tests.[5] The incidence of the disease since the programme started has been 1 in 5851, and the carrier incidence 1 in 412.[1,4] These patients are referred to their respective reference hospitals and PC teams to prevent and treat the complications of the disease.[5]

The main goal of our study was to assess, from PC, the compliance with the recommendations of the Sociedad