Academic Journal of
Pediatrics & Neonatology
ISSN 2474-7521

Juniper Publishers
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Proceeding
Volume 3 Suppl 1 – March 2017
DOI: 10.19080/AJPN.2017.03.555649

Acad J Ped Neonatol

Insulin Resistance Syndrome (Donhue)

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Submission: March 05, 2017**; Published:** March 28, 2017

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Introduction

Donohue syndrome (DS), also known as Leprechaunism, is a rare autosomal recessive form of monogenic diabetes. Affected individuals have impaired function of insulin receptor due to ballelic mutations in the coding sequence of the insulin receptor (INSR) gene. The prognosis of severe form of DS is poor and most patients die in their first year except in milder forms of the disease. This variation reflects the diversity of mutations causing the disease.

Objective

To report a 9 year old girl with severe DS and discuss the challenge of her management.

Case Report

The child presented with hyperglycaemia at the age of 45 days with classical features of DS (hypertrichosis, teeth crowding) she has no ambiguous genitalia or significant clitromegally. Initially, her blood glucose was controlled but gradually went up and insulin requirement reached up to 90 units/day. This high dose only resulted in reducing the blood glucose to an average of 200 mg/dl. In 2007 she was tried on insulin pump as her insulin requirement was very high but was stopped in 2010. Subsequently she was tried on Metformin 200 mg with no improvement. Metformin was stopped as she developed lactic acidosis. Rosiglitazone was added in 2009 with improvement of her glucose profile however mother stopped it in 2010 to avoid cardiovascular effect. Then she was started on IGF-1, which she is still on, but her HbA1c remained around 11% with fasting glucose between 238-350mg/dl.

Conclusion

This is probably one the longest survival of children with DS. The case illustrates the difficulties in the management of children with this form of diabetes.