TYPE 5 MONOGENIC DIABETES: REPORT OF 7 CASES.

INTRODUCTION:
Type 5 monogenic diabetes is an autosomal dominant disease due to a mutation in HNF1beta gene. This gene is expressed predominantly in kidney and pancreas, thus the clinical manifestations are characterised by renal abnormalities and diabetes.

METHODS:
The medical history of 7 children who were diagnosed with type 5 monogenic diabetes with genetic confirmation were reviewed.

OBJECTIVES:
To review the clinical characteristics of patients who were diagnosed with type 5 monogenic diabetes in the Pediatric Endocrinology Unit of a tertiary referral hospital.

RESULTS:
Patient information is described in the table attached.

CONCLUSION:
Phenotypic variability at the onset of Type 5 monogenic diabetes implies a diagnostic challenge.

The study of HNF1beta gene should be considered in any patient with hyperglycemia, negative antibodies for diabetes, family history of type 2 diabetes and renal abnormalities.

While progressive dysfunction of beta cells is observed, not all the patients require insulin treatment at the beginning of the disease.