



A review on specific diabetes gene and inheritance

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Background

Transient neonatal diabetes mellitus (6q24-TNDM) is related to aberrations of the imprinted gene at 6q24. The main features are severe intrauterine growth retardation, hyperglycemia that begins in infancy in a term infant and resolves by 18 months of age, dehydration, and lack of ketoacidosis. Diabetes mellitus may get worst in adolescence or later in adulthood and Women with 6q24-TNDM are at risk for being vulnerable during pregnancy.

Results

Diagnosis of 6q24-TNDM is made in a proband with transient neonatal diabetes mellitus and DNA methylation analysis that shows relative hypomethylation in the 6q24 different methylation region (DMR). 6q24-TNDM is caused by overexpression of imprinted genes at 6q24 (PLAGL1 and HYMAI). Normally, the expression of maternal alleles like PLAGL1 and HYMAI is silenced by DMR methylation, and only the paternal alleles of PLAGL1 and HYMAI are expressed. Rehydration and IV insulin is usually needed at the time of diagnosis. Subsequent relapses of diabetes may require diet modification alone, oral medications, or insulin.

Conclusion

Prevention of Secondary Complications: Prompt treatment of dehydration to prevent complications. Monitoring the growth and development of at-risk relatives: Screening for diabetes mellitus in relatives who have inherited paternal 6q24 recurrence or are at risk of inheriting two pathogenes of ZFP57.

References

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