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TOPIC: MONOGENIC DIABETES MELLITUS

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Abstract:
Aim: The main aim of the review is to describe about monogenic diabetes mellitus
Objective: Monogenic diabetes mellitus is more common in young people developing diabetes, it is important to be alert to the possibility of monogenic diabetes; hence we review about clinical features, genetics, prevalence, pathophysiology and treatment of monogenic diabetes mellitus.
Back Ground: Monogenic diabetes mellitus result from single gene mutation and account for 1-2% of cases in young people. The gene is mostly inherited from parents but may arise from spontaneous mutation. Most form of the monogenic diabetes is caused by a reduced ability to secrete insulin but rare variants result in insulin resistance.
Reason: The reason for this review is to provide broad Knowledgeof the genetic etiology of monogenic diabetes enables more appropriate treatment, better prediction of disease progression, screening of family members and genetic counselling.

Introduction:

The most common form of diabetes are type 1 and type 2, are polygenic meaning the risk of developing these form of diabetes is related to multiple genes. In contrast, rapid progress has been made over the past two decades in the identification of a range of single-gene disorders that may result in diabetes. Some of these form part of distinct genetic syndromes in which other abnormalities are prominent, and these are considered separately. The term monogenic diabetes is generally reserved for single gene disorders whose principal manifestation is diabetes, although extra-pancreatic features may also be present. Investigation of these uncommon causes of diabetes has provided considerable insight into the mechanisms underlying insulin secretion and action (1). Although monogenic diabetes may be responsible for no more than 1–2% of all cases in most populations, the diagnosis may have profound implications for treatment and prognosis, and clinicians need to be alert to this diagnosis (2).

Classification of Monogenic Diabetes:

Monogenic diabetes may arise because of defects in insulin secretion or insulin action. Insulin secretory defects account for all cases of MODY, and most cases of neonatal diabetes. Insulin resistance syndromes may be subdivided into those due to a defect in insulin signalling and those in which adipose tissue is unresponsive to insulin(1).

Mody:

MODY is a monogenic form of diabetes that usually first occurs during adolescence or early adulthood. However, MODY sometimes remains undiagnosed until later in life. A number of different gene mutations have been shown to cause MODY, all of which limit the ability of the pancreas to produce insulin(3).This process leads to the high blood glucose levels characteristic of diabetes and, in time, may damage body tis sues, particularly the eyes, kidneys, nerves, and blood vessels. MODY accounts for about 1 to 5 percent of all cases of diabetes in the United States. Family members of people with MODY are at greatly increased risk for the condition. People with MODY may have only mild or no symptoms of diabetes and their hyper glycemia may only be discovered during routine blood tests(4).MODY may be con fused with type 1 or type 2 diabetes. People with MODY are generally not overweight and do not have other risk factors for type 2 diabetes, such as high blood pressure or abnormal blood fat levels. While both type 2 diabetes and MODY can run in families, people with MODY typically have a family history of diabetes in multiple successive generations, meaning that MODY ispresent in a grandparent, a parent, and a child. Unlike people with type 1 diabetes who always require insulin, people with MODY can often be treated with oral diabetes medications. Treatment varies depending on the genetic mutation that has caused the MODY. More information about each type of MODY is provided in the appendix (1).

Neonatal Diabetes Mellitus:

NDM is a monogenic form of diabetes that occur in first 6 month of life. It is a rare condition occurring in only O'Neil 1,00,000 to 5,00,000 lives births. Infants withNDM do not produce enough insulin, lead ing to an increase in blood glucose (6). NDM can be mistaken for the much more com mon type 1 diabetes, but type 1

diabetes usually occurs later than the first 6 months of life. In about half of those with NDM, the condition is lifelong and is called permanent neonatal diabetes mellitus (PNDM). In the rest of those with NDM, the condition is transient and disappears during infancy but can reappear later in life; this type of NDM is called transient neonatal diabetes mellitus (TNDM)(7). Specific genes that can cause NDM have been identified. More information about each type of NDM is provided in the appendix. Symptoms of NDM include thirst, frequent urination, and dehydration. NDM can be diagnosed by finding elevated levels of glucose in blood or urine(8). In severe cases, the deficiency of insulin may cause the body to produce an excess of acid, resulting in a 2 potentially life-threatening condition called ketoacidosis. Most fetuses with NDM do not grow well in the womb and newborns are much smaller than those of the same gestational age, a condition called intrauterine growth restriction. After birth, some infants fail to gain weight and grow as rapidly as other infants of the same age and sex. Appropriate therapy improves and may normalize growth and development (9).

Maternally Inherited Diabetes and Diseases:

Mitochondrial diseases comprise a group of disorders produced by a failure in the oxidative phosphorilation system, the final route of mitochondrial energy metabolism, with the insulin deficiency in the biosynthesis of adenosine triphosphate (ATP) because a part of the polypeptides .The presence of maternally inherited diabetes and neurosensory deafness, to which a pattern macular dystrophy is usually added, constitute the MIDD syndrome, generally produced by a mutation of the mtDNA in position A3243 and which is responsible for 0.5%-2.8% of diabetics(10).

Conclusion:

MODY was established with the ultimate goal of improving diagnostics and treatment of monogenic diabetes. As shown by the examples discussed above, this has significantly increased our knowledge about the genetics of diabetes, and it will continue to serve scientific and diagnostic purposes in the years to come.

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