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Case Report

# Coexistence of T1DM and GCK-MODY: Case Report and Literature Review



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**What is already known on this topic?**

The coexistence of T1DM and GCK-MODY are extremely rare. When T1DM and GCK-MODY coexist, the distinctive features of GCK-MODY can be obscured due to the heightened clinical presentations of T1DM. Achieving optimal glycemic control can be particularly challenging in cases where T1DM is concomitant with GCK-MODY.

**What this study adds?**

It is crucial to recognize the complexity of diabetes types, as an individual can simultaneously have two or more different forms of diabetes. Prompt genetic testing is of particular significance, as it facilitates an early and precise diagnosis.

**Abstract**

Coexistence of Type 1 diabetes mellitus (T1DM) with glucokinase maturity-onset diabetes of the young (GCK-MODY) is extremely rare. Herein, we reported a case, conducted a systematic review and summarized the other reported cases to enhance the awareness of this rare diabetes subtype. An 11-year-old boy was presented with polydipsia, polyuria, and weight loss. He was diagnosed with T1DM based on significant hyperglycemia, decreased C-peptide levels, and positive diabetes-related antibodies. Genetic testing revealed that both the patient and his father carried a heterozygous mutation in the *GCK* gene. Due to the coexistence of GCK-MODY, the patient experienced difficulties in glycemic control and frequent hypoglycemia during insulin therapy. The patient's father gradually reduced and discontinued insulin treatment after genetic test. In clinical practice, the possibility of overlapping diabetes types should be highly emphasized. Genetic testing should be performed to optimize treatment plans and improve patient outcomes.

**Keywords:** Maturity-Onset Diabetes of the Young Type 2; Monogenic Diabetes; *GCK* Gene variation; Type 1 Diabetes Mellitus

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**Introduction**

Maturity-Onset Diabetes of the Young (MODY) is a monogenic diabetes characterized by autosomal dominant inheritance. It arises from gene mutations that impair β-cell function, pancreatic development, and insulin secretion, ultimately causing hyperglycemia(1). Its prevalence is estimated to range from 1% to 6% among children with diabetes(2). The characteristics of MODY mainly include onset during adolescence (with an age of onset ranging from 6 months to 25 years), absence of diabetes-related autoantibodies, non-insulin dependency, and often a family history of diabetes.

GCK-MODY (also known as MODY2) was first described in 1992 by Froguel, P. (3) and colleagues in France. It is one of the common subtypes of MODY, accounting for approximately 10% to 60% of MODY cases(4). The prevalence of GCK-MODY exhibits ethnic and geographic heterogeneity. A multicenter real-world study revealed that GCK-MODY accounted for 57.4% of MODY cases in Germany(5). Conversely, data from a cohort study in Norway indicated that GCK-MODY constituted 12%(6). GCK-MODY is predominantly characterized by chronic, non-progressive elevated fasting blood glucose. This condition is well-managed through dietary control, and pharmacotherapy is rarely required. Additionally, the incidence of diabetic microvascular and macrovascular complications is exceedingly rare. GCK-MODY is caused by heterozygous inactivating mutations in the glucokinase (GCK) gene. Glucokinase, a key enzyme in glucose metabolism, plays a crucial role in pancreatic β-cell function. Impaired glucokinase function results in reduced sensitivity of β cells to glucose, thereby increasing the glycemic threshold required for insulin secretion. This alteration leads to a decrease in insulin secretion, which ultimately culminates in hyperglycemia.

Type 1 diabetes mellitus (T1DM) accounts for 5-10% of the diabetes and is characterised by a complete insulin deficiency due to the autoimmune destruction of pancreatic β-cells(7). T1DM typically present with polydipsia, polyphagia, polyuria, and weight loss. Diabetic ketoacidosis (DKA) occurs in approximately 41.7% of children when they are first diagnosed with T1DM(8). Patients with T1DM require lifelong insulin replacement therapy.

As the low prevalence of both T1DM and GCK-MODY, the coexistence of the two diabetes types is extremely rare. To date, only four cases have been reported. Overlap of these two diabetes types presents with unique clinical manifestations. In this study, we report a child with T1DM and GCK-MODY who was admitted to our hospital in July 2024. Additionally, we conducted a systematic review and summarized the other reported cases to enhance the awareness of this rare diabetes subtype.

**Case Report**

An 11-year-old boy was presented to our clinic with complaints of polydipsia, polyphagia, and polyuria for the past two weeks. His fasting blood glucose was 14.2 mmol/L. The patient had no fever and no significant past medical history. On physical examination, the patient was 139cm in height and weighed 27kg, with a body mass index (BMI) of 14 kg/m² (z score: -2.2). Acanthosis nigricans was not presented. Birth weight was 3200 g. The patient's mother had a history of gestational diabetes. The patient's father was diagnosed with type 2 diabetes mellitus at the age of 30. His precise anthropometric data, including weight, height, and BMI, at the time of diagnosis is unavailable. However, he reported being underweight at that time. At the age of 34, he initiated treatment with acarbose and insulin. The father's diabetes-related autoantibodies were all negative. At the age of 37, the patient's father underwent a standard meal test, and the results are shown in Figure 1.

Glycated hemoglobin (HbA1c) of our case was 11.7% (reference range: 4-6%). His fasting blood glucose was 15.15 mmol/L, fasting C-peptide was 0.37 ng/mL (reference range: 0.4-5.7 ng/mL), and fasting insulin was 2.3 uU/mL (reference range: 1.9-23 uU/mL). Urinalysis showed 3+ glucosuria and 2+ ketonuria. Anti-glutamic acid decarboxylase antibodies (GADA) were >2000 IU/mL (reference range: <10 IU/mL), IA-2A antibodies was 77.11 IU/mL (reference range: <10 IU/mL), and anti-islet cell antibodies (ICA) was 43.75 COI (reference range: <1 COI). Anti-insulin antibodies (IAA) and zinc transporter 8(ZnT8) antibodies were both negative. The patient's cholesterol, triglycerides, low-density lipoprotein (LDL), and high-density lipoprotein (HDL) levels were all within the normal range. Thyroid function, cortisol levels, and adrenocorticotropic hormone (ACTH) levels were also normal. Antinuclear antibodies (ANA) were negative. One month after initiating insulin therapy, the patient underwent a standard meal test, the results are shown in Figure 1.

After obtaining informed consent, 3 mL of peripheral blood samples anticoagulated with ethylenediaminetetraacetic acid (EDTA) were collected from the patient and his parents. The whole-exome sequencing results revealed a heterozygous c.751A>C mutation in exon 7 in the *GCK* gene. This mutation is a missense that leads to an amino acid change from methionine to leucine at position 251 (p. Met251Leu). This mutation has a frequency of 0 in the general population database. It has been reported in patients with GCK-MODY in the literature and is classified as a pathogenic mutation. Additionally, pathogenic variants with different amino acid substitutions at the same codon have been documented. The REVEL protein function prediction tool classified this mutation as deleterious. Sanger sequencing validation demonstrated that the patient's father carried the same heterozygous variant at this locus, while the patient's mother had a normal genotype at this site.

The patient was diagnosed with T1DM in combination with GCK-MODY and was treated with insulin at a dosage of 0.66 U/kg/d. Three months after treatment, the HbA1c decreased to 6.9%. The fasting blood glucose was maintained between 5.0 and 7.0 mmol/L, and the 2-hour postprandial blood glucose ranged from 4.5 to 9.0 mmol/L.

**Discussion**

We present a pediatric case affected by two types of diabetes, T1DM and GCK-MODY. The patient exhibited typical symptoms of polydipsia, polyuria, and weight loss. A markedly elevated blood glucose level, weak insulin production, and positive diabetes autoantibodies support the diagnosis of T1DM. Given that the father was diagnosed with T2DM at the age of 30s, further genetic testing revealed that both our patient and father carried a heterozygous mutation in the *GCK* gene. The diagnosis of GCK-MODY is definitive. Despite the presence of an identical *GCK* gene mutation in both the child and the father, their clinical phenotypes exhibited notable heterogeneity. The child and the father exhibited differential susceptibility to autoimmune diabetes, which is presumably attributable to their distinct HLA genotypes(9).

The coexistence of T1DM and GCK-MODY is extremely rare, with only four cases reported to date (Table 1). Both Case 1 and 3 presented with the typical clinical manifestations of T1DM. As these cases have the presence of dysglycemia in first- or second-degree relatives, further refinement of the gene sequencing revealed a combination of GCK-MODY, which was the same as in our case. The symptoms associated with T1DM are frequently more pronounced in comparison to those observed in MODY, and the co-occurrence of both conditions has been documented. Therefore, it is recommended that children diagnosed with T1DM who have a family history of diabetes undergo early genetic evaluation. This enables the timely identification of coexisting monogenic diabetes.

In addition, the father of the child in this study was diagnosed with type 2 diabetes mellitus at the age of 30, starting treatment with oral hypoglycemic agents and insulin at the age of 34. He was diagnosed with GCK-MODY at the age of 43, following genetic testing prompted by his child's diagnosis. After diagnosis he stopped insulin therapy and started dietary glycemic control instead. In clinical practice, MODY is often misdiagnosed as type 1 or type 2 diabetes mellitus and treated as such in the long term. Studies have shown that approximately 80% of MODY patients remain undiagnosed(10). Therefore, patients suspected of having monogenic diabetes mellitus should undergo genetic testing to develop a precise treatment plan. The high cost of genetic testing and the lack of understanding of the genetic profile of monogenic diabetes and the clinical screening process are challenges(11). Current clinical screening strategies include the GCK-MODY clinical screening strategy(12), as well as the MODY Calculator（https://www.diabetesgenes.org）. These screening strategies were not precise enough when used in cases like the pediatric patients in our study. The results of the MODY calculator for our case showed that there was only a 2.6% probability of determining a MODY. Future research should focus on developing more scientific and precise clinical screening methods to improve the cost-effectiveness and accuracy of genetic screening for monogenic diabetes.

Diabetes is increasingly recognized as a heterogeneous metabolic disorder. Although T1DM, T2DM, and MODY exhibit distinct pathophysiological mechanisms, their clinical presentations frequently overlap. Some patients demonstrate characteristics consistent with two or more diabetes types concurrently. For instance T1DM overlaps with MODY, T1DM complicated by metabolic syndrome (also known as Double Diabetes)(13)，overlap between T2DM and MODY (14), etc. An individual can experience different types of diabetes simultaneously or successively. In 2012, Calcaterra V et al. reported a 7-year-old girl from Italy diagnosed with GCK-MODY. She gradually developed features of obesity and metabolic syndrome at the age of 12, and then her antibody status converting from negative to positive and her antibody titers progressively increasing. She was eventually diagnosed successively with GCK-MODY, metabolic syndrome and T1DM(15).

According to the literature, T1DM and GCK-MODY can occur either sequentially or simultaneously. Initial diagnoses of both Case 1(16) and Case 3(17) were T1DM (positive autoantibodies, insulin dependence).

Subsequent genetic testing, prompted by family history, revealed concurrent GCK-MODY. Case 2(18) and Case 4(19)initially diagnosed as GCK-MODY (mild fasting hyperglycemia, pathogenic *GCK* gene variants). A subsequent development of T1DM was observed, marked by progressive glucose fluctuations and sustained elevation of diabetes autoantibodies (as confirmed by laboratory findings in Table 1). A comprehensive genetic evaluation is necessary even after a diagnosis of autoimmune diabetes, particularly in patients exhibiting atypical glycemic patterns or a strong family history. For patients with GCK-MODY, clinicians must maintain a high level of vigilance for substantial glycemic fluctuations throughout their lifetime. When such changes occur, a prompt reassessment of diabetes classification is imperative.

T1DM coexisting with other MODY types is extremely rare. Only two cases have been reported (Table 2). Both cases exhibit highly complex disease trajectories, underscoring the fact that diabetes is a heterogeneous and profoundly individualized condition. One patient was initially diagnosed with T2DM. Due to a strong family history (the father, paternal aunt and uncle, and grandmother were all diagnosed with T2DM between ages 40–50). Genetic testing was later performed and revealed *HNF-1α* gene mutation. Despite multiple oral hypoglycemic agents, the patient's glycemic control deteriorated, accompanied by significant weight loss and positive diabetes autoantibodies, casting doubt on the original T2DM diagnosis. Notably, the patient lacked clinical signs of insulin resistance (e.g., acanthosis nigricans). Given the patient's obesity, positive autoantibodies, family history, and genetic findings, the authors concluded that the most accurate diagnosis was "triple diabetes," which is the coexistence of obesity, islet autoimmunity, and HNF1A-MODY. The authors hypothesized that metabolic syndrome and insulin resistance, both of which are triggered by obesity, may have caused β-cell destruction and dysfunction, ultimately resulting in progressive insulin deficiency(20). Another patient was diagnosed with T2DM at age 30. After 20 years of metformin treatment, insulin therapy was initiated. The patient's mother and multiple maternal second-degree relatives have been documented as having diabetes. Genetic testing revealed a pathogenic mutation in the *HNF-1α* gene (c.872dupC, p. Gly292Argfs*25). At age 68, anti-glutamic acid decarboxylase antibodies (GADA) were incidentally detected, suggesting a coexistence of T1DM and HNF1A-MODY(21).

It has been demonstrated that patients diagnosed with GCK-MODY do not require treatment and have a favorable long-term prognosis(5,22). However, the treatment strategy for patients with concomitant types of diabetes becomes more complex. In cases where T1DM is concomitant with GCK-MODY, achieving optimal glycemic control can be particularly challenging. A low dose of insulin has been demonstrated to be ineffective in controlling glucose levels. Conversely, a high dose of insulin has been shown to result in hypoglycemia. In the present study, the child, as well as Case 1 and Case 3, exhibited recurrent hypoglycemia during insulin therapy and demonstrated difficulty in achieving target glycated hemoglobin levels(16,17). Mutations in the *GCK* gene may cause impairment of counter-regulatory response to hypoglycemia(23). Previous studies have shown that GCK plays an important role in the ability to sense hypoglycemia in pancreatic α cells(24). Chakera et al. conducted a hyper insulinemic-euglycemic glucose clamp study in patients with GCK-MODY, individuals with T2DM, and healthy controls. The results demonstrated that, during progressive hypoglycemia, patients with GCK-MODY exhibited elevated thresholds for glucose production(25). For patients diagnosed with both T1DM and GCK-MODY, the administration of high-dose insulin to reduce blood glucose levels may result in recurrent hypoglycemia, thereby complicating disease management. Therefore, Aoife Garrahy et al. proposed that the target blood glucose for insulin therapy should be set at a fasting blood glucose of 7–8 mmol/L(16).

Compared with T1DM, patients with overlapping T1DM and T2DM reveals a significantly high risk of developing microvascular and macrovascular complications(26). Currently, reports on the coexistence of T1DM and GCK-MODY are extremely rare. Therefore, the potential for microvascular and macrovascular complications in this limited patient population in the future remains uncertain, necessitating ongoing long-term observation.

**Conclusion**

In cases when T1DM and GCK-MODY coexist, the distinctive features of GCK-MODY can be obscured due to the heightened clinical presentations of T1DM, as T1DM is accompanied by a substantial decrease in insulin C-peptide levels and notable abnormalities in blood glucose levels. Consequently, prompt genetic testing is of particular significance, as it facilitates an early and precise diagnosis. In clinical practice, it is crucial to recognize the complexity of diabetes types, as an individual can simultaneously have two or more different forms of diabetes. Achieving optimal glycemic control can be particularly challenging in cases where T1DM is concomitant with GCK-MODY. *GCK* gene mutation reduces sensitivity to blood glucose levels, which makes controlling glycemia with exogenous insulin difficult and predisposes individuals to recurrent

hypoglycemia.

**Conflict of interests:** The authors declare that they have no competing interests.

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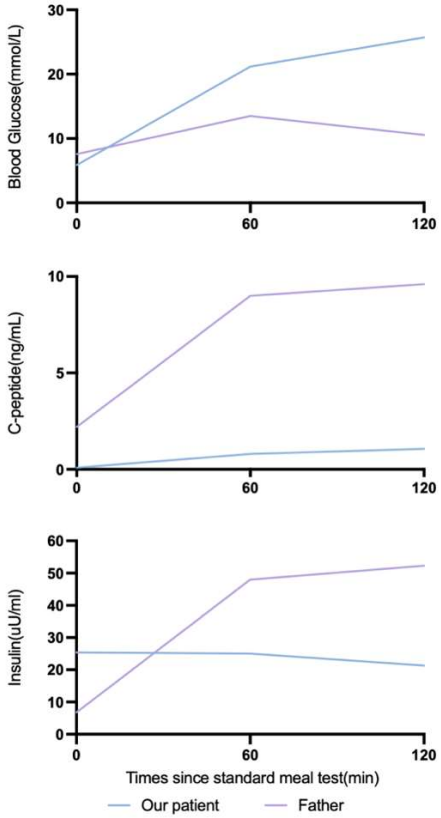


**Figure 1.** Two hours standard meal test

| **Table 1. Characteristics of patient diagnosed T1DM and GCK-MODY** | | | | | |
|---|---|---|---|---|---|
| | Our Case | Case 1 | Case 2 | Case 3 | Case 4 |
| Country | China | Ireland | Switzerland | England | Italy |
| Age of onset | 11y | 32y | 10y | 14y | 4y |
| Gender | M | F | M | M | M |

| | | | | | |
|---|---|---|---|---|---|
| *GCK* mutation | c.751A>C | c.478G>C | C.1305-1306INSG | heterozygous deletion on exons 5 and 6 | c.863+2T>A |
| Family history | Father: diabetes (diagnosed at 31)<br>Mother: gestational diabetes | Cousin: MODY2 (diagnosed) | Sister: MODY2 (diagnosed) | Elder brother and grandfather: MODY2 (diagnosed) | Father, elder sister, younger brother: MODY2 (diagnosed) |
| BMI（$kg/m^2$） | 13.8 | 21.6 | NA | NA | 15.9 |
| HbA1c(%) | 11.74 | 13.60 | 12.85 | 13 | 6.40 |
| Fasting Blood Glucose（mmol/L） | 15.15 | 20.3 | 29 | 23 | 11.1 |
| Fasting c-peptide（ng/ml） | 0.37 | 0.16 | 0.26 | NA | 0.3 |
| 2h c-peptide（ng/ml） | 1.06 | NA | 0.42 | NA | 1.2 |
| Autoantibody | GADA、IA-2A、ICA positive | GADA positive | GADA、IA-2A positive | GADA positive | GADA、ICA positive |
| Insulin（U/kg/d） | 0.66 | 0.63-0.85 | NA | 0.75–0.85 | 0.7-1.0 |
| HbA1c after treatment | 6.90 | 7.34 | NA | 7.50 | 6.70 |
| NA: Not Available | | | | | |

**Table 2. Characteristics of patient diagnosed T1DM and HNF1A-MODY**

| | Case 1 | Case 2 |
|---|---|---|
| Country | America | Switzerland |
| Age of onset | 17y | 73y |
| Gender | F | M |
| *HNF-1α* mutation | NA | c.872dupC |
| Family history | Father, father's siblings, grandmother: T2DM (diagnosed) | Mother and maternal second-degree relatives: DM (diagnosed) |

| | | |
|---|---|---|
| BMI （$kg/m^2$） | 36.4 | 27 |
| HbA1c (%) | 11.90 | 7.30 |
| Fasting Blood Glucose（mmol/L） | 13 | NA |
| Fasting c-peptide（ng/ml） | 3.3 | 0.22 |
| 2h c-peptide（ng/ml） | NA | 0.33 |
| Autoantibody | GADA、ICA positive | GADA positive |
| Insulin（U/kg/d） | 0.6 | 0.62 |
| HbA1c (%) | NA | 6.60 |
| NA: Not Available | | |