International Journal of Contemporary Pediatrics
Syal S et al. Int J Contemp Pediatr. 2026 Jan;13(1):95-98
http://www.ijpediatrics.com
pISSN 2349-3283 | eISSN 2349-3291

DOI: https://dx.doi.org/10.18203/2349-3291.ijcp20254193

Case Report

Cataract in childhood diabetes: more than what meets the eye

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Received: 20 November 2025
Accepted: 18 December 2025

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ABSTRACT

Diabetic cataract is a rare but sight threatening complication in children. Two cases of paediatric diabetic cataract are described, one is in a six-year-old girl with diabetes duration of four years with systemic features of poorly controlled T1DM in the form of Mauriac syndrome and another is of a 13 years 6 months old boy with relatively recent diabetes diagnosis 6 months back, both presenting with visually significant cataracts. Regular ophthalmological screening examinations in patients with T1DM starting right from the time of diagnosis are essential to pick up and treat this complication early for best results.

Keywords: Type 1 diabetes mellitus, Cataract, Mauriac syndrome, Ophthalmological screening

INTRODUCTION

Diabetes mellitus (DM) is a common chronic metabolic disease characterized by hyperglycemia as a cardinal biochemical feature.[1] The major forms of diabetes are differentiated by insulin deficiency versus insulin resistance: type 1 diabetes mellitus (T1DM) results from deficiency of insulin secretion because of pancreatic β-cell damage whereas type 2 diabetes mellitus (T2DM) is a consequence of insulin resistance occurring at the level of skeletal muscle, liver and adipose tissue, with various degrees of β-cell impairment. T1DM is the most common endocrine metabolic disorder of childhood and adolescence with important consequences for physical and emotional development. This disorder is accompanied by various complications; both in the short term and long term. Acute complications include diabetic ketoacidosis and hypoglycemia whereas chronic microvascular complications include diabetic nephropathy, retinopathy and neuropathy. Ocular complications are rarely discussed in paediatric diabetes as they are a result of chronic long-term hyperglycemia and not very commonly seen in paediatric age group. Most ophthalmological emphasis has been laid on diabetic retinopathy (DR). Diabetes, however, is also a risk factor for the development of cataracts which are one of the significant causes of visual impairment. Diabetic cataracts are mostly seen in young adults due to prolonged poor glycemic control. In the paediatric age group, these are usually seen in adolescents with disease onset at an early age.

Two cases of T1DM presented with significant visual disturbances due to cataract. The first patient was a 6-year-old female which is one of the youngest in literature to develop diabetic cataract. The first patient had T1DM since last 4 years with systemic features suggestive of Mauriac syndrome. The second patient was an adolescent male who had cataract as an early ocular complication as the case had been diagnosed with T1DM just 6 months back. These cases highlight the spectrum of presentations of pediatric diabetic cataract, underscoring the need for vigilance in both recently diagnosed and long standing T1DM (Table 1).

Table 1: Comparison of the two pediatric diabetic cataract cases.

Characteristics	Case 1	Case 2
Age/sex	6 year old female	13 yrs 6 months old male
Duration of T1DM	4 years	6 months
Compliance with Insulin therapy	Poor	Poor
Ocular findings	Bilateral cataract changes seen on slit lamp. Fundus not visualized due to insufficient optical clarity owing to presence of cataractous lens	Bilateral corneal opacities visible to the naked eye. Slit lamp showed bilateral mature cataract with non-visualization of posterior pole. Fundus view not possible due to insufficient optical clarity because of the cataractous lens.
Systemic findings	Growth retardation, palpable liver	Normal
Other diabetic complications, except cataract	Probable Mauriac syndrome	Nil

CASE REPORT

Case one is a 6-year-old female, known case of T1DM, who presented with diminution of vision in both eyes since the last 2 months which was gradual, painless and progressive. The first patient had difficulty in reading the blackboard impairing her scholastic performance. No history of floaters, flashes of light or dark spots. No history of trauma, redness, pain or discharge from eyes. The first patient was diagnosed with T1DM at 2 years of age when she had complaints of increased frequency of urination and increased thirst and was started on Mixtard insulin regimen, presently having poor glycemic control. She was first born of a non-consanguineous marriage with normal perinatal transition and development. On examination, her weight and height were at 3rd centile (within midparental height range). The first patient was normotensive, prepubertal and had hepatomegaly with a liver span of 11 cm, firm and non-tender. Ocular examination showed bilateral cataract changes on slit lamp evaluation (Figure 1).

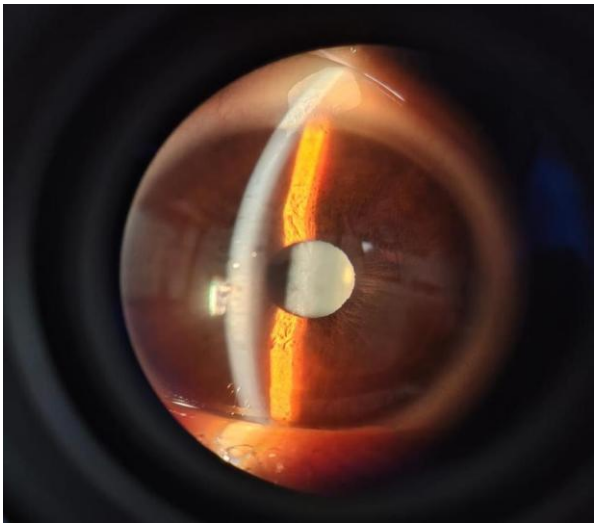

Figure 1: Slit lamp examination showing posterior subcapsular cataract.

Fundus view was not possible due to media haze (Figure 2). Based on the clinical profile, the probability of Mauriac syndrome was considered. Investigations revealed microcytic hypochromic anaemia with normal liver and kidney function tests. Her HbA1c was 18.4 % with normal thyroid profile and celiac screen. Anti Glutamic acid decarboxylase (GAD) 65 Antibody was positive. Fasting lipid profile and urine microalbumin: creatinine were normal. Child was shifted to basal bolus regimen of insulin with titration of doses to achieve euglycemia. Given the presence of a dense, visually significant posterior subcapsular cataract obscuring the visual axis, cataract extraction has been planned.

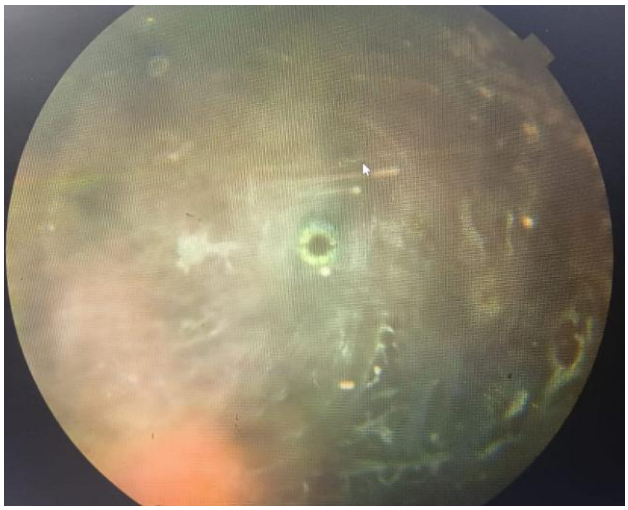

Figure 2: Fundus photograph showing diffuse media haze with poor visualization of the optic disc and macula, consistent with significant lens opacity, in a child with T1DM.

Case two is a 13 year 6 months old male, known case of T1DM diagnosed 6 months back when presented with polyuria, polydipsia and weight loss and was started on subcutaneous insulin therapy. The case now presented with blurring of vision in both eyes since the last one

month which was painless and progressive and was impairing the child's ability to take insulin, thus, deteriorating his glycemic control. The case was third born of a non-consanguineous marriage with normal perinatal transition and development. On examination, his weight was at 10th centile and height at 25th centile within midparental height range. Eye examination showed bilateral corneal opacities. Slit lamp examination revealed bilateral mature cataract with non-visualization of posterior pole. Fundus could not be visualized due to significantly hazy media. His Sexual Maturity Rating was tanner stage III with rest of the general and systemic examination showing no abnormalities. Investigations revealed normal hemogram, liver and kidney function tests. His HbA1c was 15.9% with normal thyroid profile, celiac screen and fasting lipid profile. After achieving euglycemia with titration of insulin doses, the child underwent bilateral phacoemulsification with intraocular lens implantation with good visual outcome.

DISCUSSION

T1DM is caused by autoimmune destruction of beta cells of pancreas leading to chronic hyperglycemia and contributing significantly to childhood morbidity and mortality. Approximately 8.5 million individuals in the world have T1DM with an estimation of 80,000 new cases every year.[1] Ocular complications are rarely discussed in paediatric diabetes as they are a result of chronic long-term hyperglycemia and not very commonly seen in paediatric age group. However, when they do occur, they contribute significantly to morbidity in these patients by impairing vision, especially in this critical period of knowledge acquisition and learning. It also impairs the glycemic control especially in older children who are self-administering insulin and thus contributes to a vicious cycle.

Most ophthalmological attention has been paid to the screening, diagnosis and management of diabetic retinopathy. However, it is important to remember that cataract in diabetes mellitus is also a significant contributor to visual morbidity and untreated cataracts in children cast tremendous burden to the child and family.

The peak incidence of diabetic cataract is seen at the age of 20-29 years.[2] In paediatric age group, these are usually seen in adolescence in children who were diagnosed with T1DM at an early age as a result of long-term hyperglycemia. However, early cataracts have also been described with an incidence of 0.7-3.4% which can occur as early as at the time of diagnosis of T1DM with the youngest patient being reported at the age of 5 years.[3-5]

The pathogenesis of diabetic cataract is multifactorial. Aldose reductase, an enzyme involved in the alternative glucose metabolic pathway, primarily resides in the lens epithelium and converts excess glucose to sorbitol when blood glucose levels are elevated. Sorbitol accumulation increases the osmotic pressure of lens cells, leading to vacuolization in the lens, lamellar separation and eventually development of cataract.[6] However, the occurrence of early diabetic cataracts suggest that other pathophysiological mechanisms are also implicated. Acute osmotic changes in the lens epithelium due to fluctuating glucose, oxidative stress due to reduced glutathione stores and autoimmune factors have also been proposed to be contributory.[7,8]

Wilson et al. described pediatric diabetic cataracts of varied morphology from posterior subcapsular, snowstorm, lamellar, flake-like and dense milky-white cataract with most common location being posterior subcapsular pole.[4] The variation in cataract morphology in these children may be explained by differences in age of DM diagnosis and severity of disease. These progress rapidly over the course of weeks to months causing accelerated visual loss. Surgical extraction is the current gold standard in the management of early, visually impairing cataracts in patients with diabetes. Though, there have been isolated reports of regression of metabolic cataracts with good glycaemic control, but there is stronger evidence to support early surgical management, especially among those with poorer visual acuity.[3]

Currently, most guidelines including American Diabetes Association (ADA) and American Academy of Paediatrics (AAP) recommend screening for chronic ocular complications in T1DM to begin 3-5 years after diabetes onset of puberty or age 11 years.[9,10] However, these recommendations largely pertain to diabetic retinopathy as it is considered to take at least 3-5 years to develop after the onset of hyperglycemia. The cases showing early and accelerated development of cataract in T1DM is a good reminder to consider early ocular screening at the time of diagnosis in all diabetic children, which has also been recommended by International Society for Pediatric and Adolescent Diabetes (ISPAD).[11] Hence, it is imperative to get regular ophthalmological examinations starting right from the time of diagnosis in patients with T1DM so that this complication can be picked up and treated early for best results.

CONCLUSION

Pediatric diabetic cataract remains a rare but important cause of preventable visual loss. These cases highlight that cataracts may present both early and late in the disease course, sometimes with systemic manifestations such as Mauriac syndrome. Prompt recognition, multidisciplinary care and timely treatment with appropriate visual rehabilitation are essential to preserve vision and quality of life.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

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Cite this article as: Syal S, Goel AK, Jagzape TB, Khan L, Jaiswal MD, Gandhoke CS. Cataract in childhood diabetes: more than what meets the eye. Int J Contemp Pediatr 2026;13:95-8.