Dexmedetomidine-based intravenous sedation of a Glucose-6-phosphate dehydrogenase deficiency pediatric patient: A case report

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Introduction

Glucose-6-phosphate dehydrogenase (G6PD) deficiency is a very common X-linked genetic disorder caused by a structural abnormality in the G6PD enzyme. The G6PD enzyme catalyzes the first step in the pentose phosphate pathway, leading to antioxidants that protect red-blood-cells against oxidative damage. (Fig. 1)G6PD deficiency can cause hemolytic anemia, usually after exposure to certain medications, foods, and infections. Therefore, clinical management of G6PD deficiency is to prevent hemolysis caused by oxidant stress from certain drugs, and severe infections. To the best of our knowledge, **this is the first reported case of dexmedetomidine-based intravenous sedation used in a G6PD deficiency patient.**

Patient and Methods

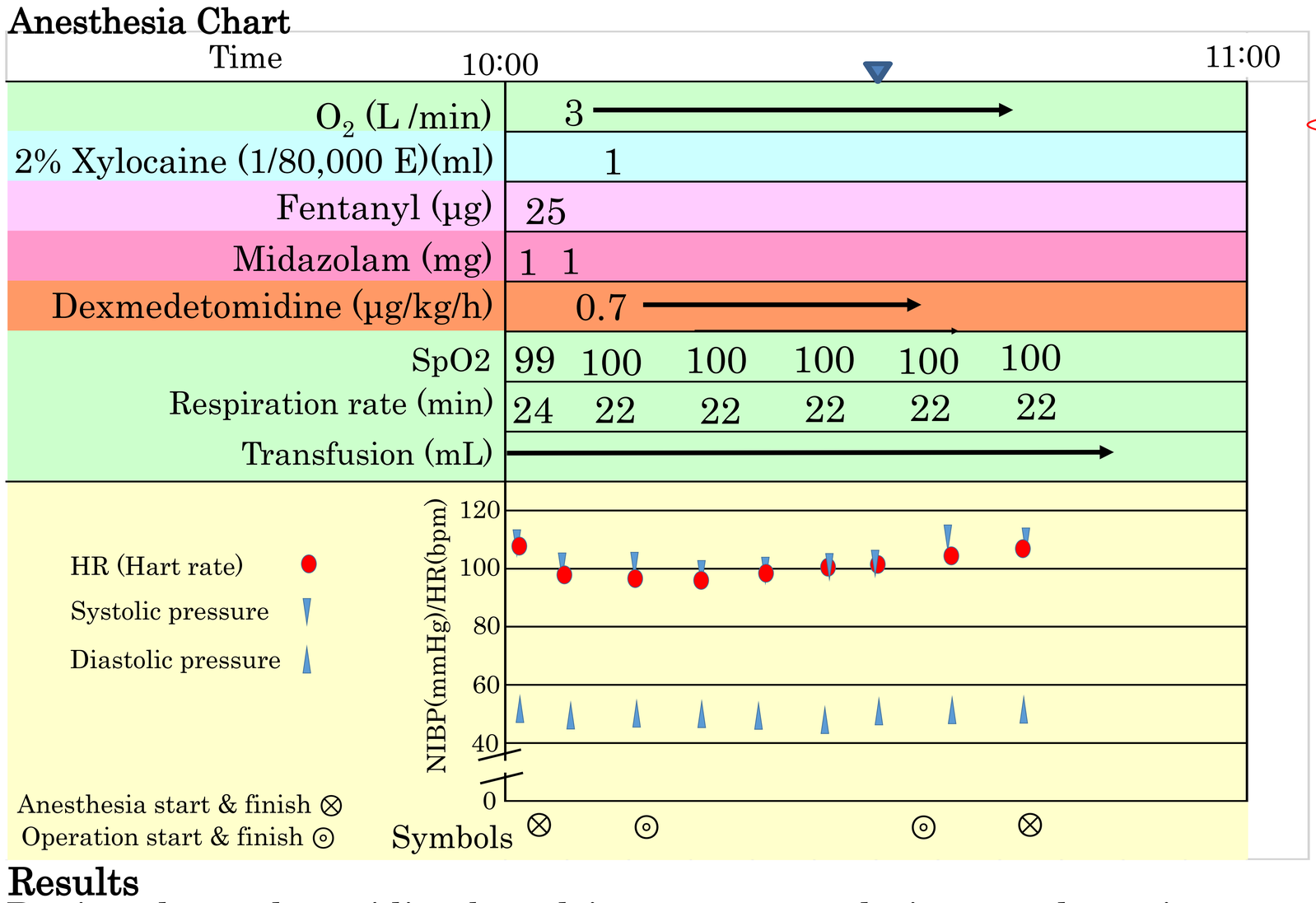
Patient

The patient was a 5-year-old boy (height 115 cm, body weight 22 kg) with G6PD deficiency. (Fig. 2.) He had no previous medical history of hemolytic anemia. He did not have any problems when he took painkillers or cold medicines. The patient's grandfather, who was of Taiwanese origin, also had G6PD deficiency; however, the grandfather also had never developed a hemolytic reaction. (Fig. 3) The patient had mild amblyopia and a mild mental retardation. We performed frenectomy under intravenous sedation.

Methods

We used dexmedetomidine-based intravenous sedation. Dexmedetomidine has been reported to have antioxidant activity, to cause less respiratory depression than other sedatives, and to be effective for pediatric sedation. And, we used other sedative drugs before using dexmedetomidine, to avoid a change in circulation at the time of dexmedetomidine loading. His vital signs were stable and maintained a Ramsay Score of 4.

Anesthesia Chart



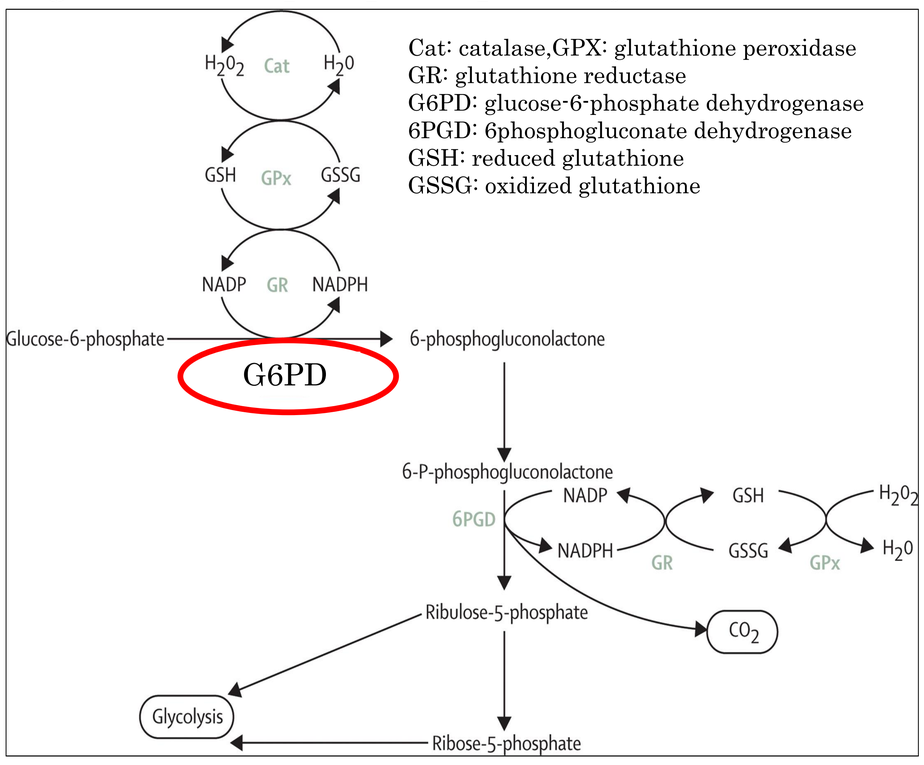
Results

During dexmedetomidine-based intravenous sedation, good respiratory and circulatory states were maintained. No problems occurred in the perioperative period; signs of hemolysis such as fatigue, headache, and dark urine were not observed.

Conclusions

Our case suggests that the selection and use of sedatives with anti-oxidant and anti-inflammatory effects to counter the rise in perioperative oxidative stress will increase safety. Dexmedetomidine was safe and effective for this pediatric patient with G6PD deficiency. We suggest that dexmedetomidine is a safe drugs that can be used for pediatric patients with G6PD deficiency.

Fig. 1. Pentose-phosphate pathway



Lancet 2008; 371: 64-74

Fig. 2. Patient's G6PD enzyme data

Normal range (mean ± SD)	Control	Our patient
(7.61–9.81) (IU/gHb)	10.2	0.7

Fig. 3.Patient's family

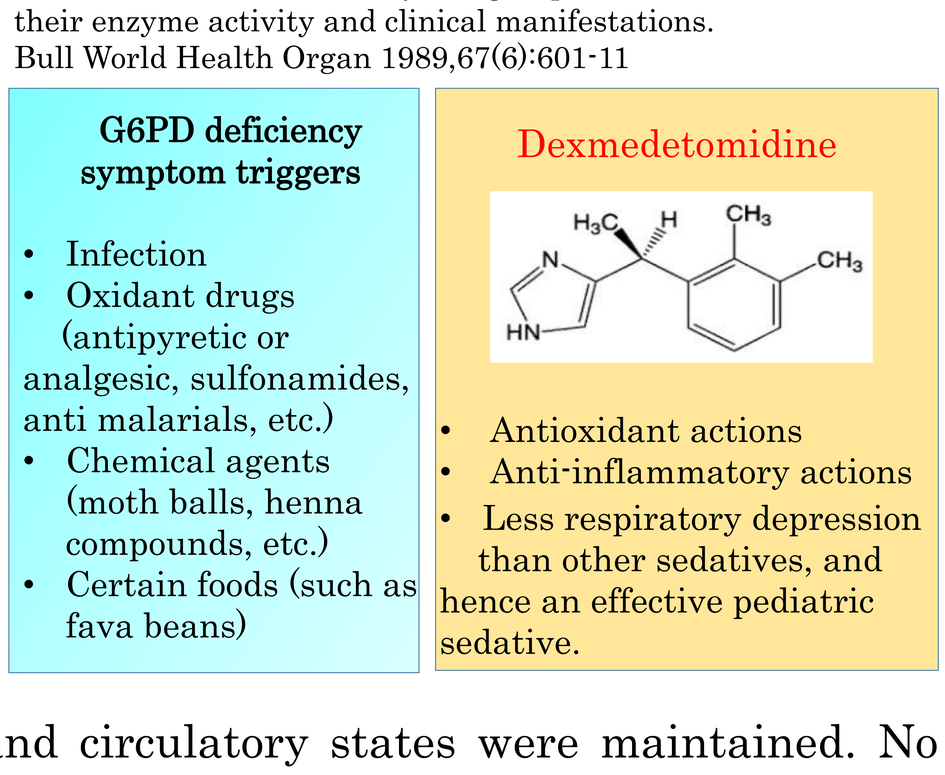


Fig. 4. WHO classification of G6PD deficiency

Class Ⅰ	Severely deficient chronic hemolytic anemia
Class Ⅱ	1%-10% residual activity ← Our patient
Class Ⅲ	10%-60% residual activity
Class Ⅳ	60%-150% normal activity
Class Ⅴ	>150% increased activity

Variants of G6PD deficiency are grouped into 5 classes based on their enzyme activity and clinical manifestations.
Bull World Health Organ 1989,67(6):601-11

G6PD deficiency symptom triggers

- Infection
- Oxidant drugs (antipyretic or analgesic, sulfonamides, anti malarials, etc.)
- Chemical agents (moth balls, henna compounds, etc.)
- Certain foods (such as fava beans)

Dexmedetomidine

- Antioxidant actions
- Anti-inflammatory actions
- Less respiratory depression than other sedatives, and hence an effective pediatric sedative.

References

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