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Título	Efeito a Curto Prazo da Empagliflozina em Pacientes com Síndrome de Berardinelli
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Short Term Effect of Empagliflozin in Patients with Seip-Berardinelli Syndrome

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Resumo:

Seip-Berardinelli Syndrome or congenital generalized lipodystrophy is characterised by the lack of adipose tissue, with its consequent deregulations in lipid and carbohydrate metabolism leading to, among other conditions, an early on set, hard to manage, insulin resistant diabetes. There are very few articles in the literature approaching the treatment for diabetes in these patients. Our experience shows that even with metformin and a controlled diet this diabetes can be very hard to manage. Thus in order to improve symptoms and laboratory indicators of chronic hyperglycemia we associated empagliflozin to all of our Seip-Berardinelli patients who had already developed a hard-to-manage diabetes. After empiric use of empagliflozin in one patient, with impressive improvement in laboratory tests, we started prescribing the drug to all of our Seip-Berardinelli patients who had already developed a hard-to-manage diabetes. We analyzed fasting blood glucose levels, glycosylated hemoglobin, triglycerides, total cholesterol, HDL cholesterol, LDL cholesterol, C-peptide and blood insulin levels of all four patients before the start of the drug and 3 months after. All patients were advised to continue with their low carb and low fat diet. We prescribed 10mg of empagliflozin once a day in addition to the other medication they had been using. All patients had a significant reduction of fasting blood glucose levels of -96mg/dL (CI 59,9 to 132.1; $p=0,003$). The change in glycosilated hemoglobin was of $-1,575\%$ (CI $-0,45$ to $3,7$; $p=0,09$). We speculate that one of the reasons for the immediate success of empagliflozin compared to other oral hypoglycemic agents in Berardinelli patients is that it acts independently of insulin. The follow-up of these patients is important to elucidate the long term effect of empagliflozin as well as its possible side benefits in this particular syndrome. But we believe, based on our short term effect serial case, that the use of this drug might be a turning point on managing diabetes in Berardinelli-seip patients.