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Hyperpigmentation in Only One Identical Twin

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A 10 year-old previously healthy female presented to the hospital in the winter season with vomiting and lethargy. Laboratory studies showed hypoglycemia (42 mg/dl), hyponatremia (126 mmol/l), hyperkalemia (5.7 mmol/l), and acidosis (bicarbonate 12 mmol/l). She was also hypotensive with a Doppler blood pressure of 68/22. She subsequently recovered with intravenous fluid hydration and glucose. She tested positive for influenza A, and she was discharged home with a suspected viral illness. She returned to the hospital exactly one month later with similar symptoms, and on examination it was noted that her skin was now tan when compared to her identical twin sister (Picture). She also had a hyperpigmented scar on her right knee and hyperpigmentation of her umbilicus (Picture). Her baseline cortisol level was 1.3 µg/dl and rose to 1.8 µg/dl after a high dose ACTH (250 µg) stimulation test. Additional testing revealed primary autoimmune adrenal insufficiency (Addison's disease). Her symptoms improved after starting hydrocortisone and fludrocortisone. Nearly four years later, her identical twin remains disease free.