

## Barraquer Simons Syndrome - Rare Cause of Progressive Wasting in an Eight Year Old Girl

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### ABSTRACT

Partial Lipodystrophy is rare cause of progressive wasting in children. This article describes a child who had progressive wasting of the upper half of the body who was diagnosed to have acquired partial lipodystrophy. Knowledge of this rare disorder would be useful while evaluating a child with chronic wasting.

An eight-year-old girl, the first born child out of a non-consanguineous marriage, was brought with a history of progressive thinning of face gradually progressing to involve the body, beginning from six years of age, despite adequate nutritional intake (Figure 1a and 1b). There was no history of prolonged fever, loss of appetite, steatorrhea, recurrent vomiting, polyuria, polydipsia and recurrent infections needing hospitalizations.

On clinical examination, child was thin built, with thinning of the face and upper half of the body (Figure 2a-c). The lower limbs were relatively spared. There was no acanthosis nigricans. Blood pressure was normal. There was no pallor or generalized lymphadenopathy. Systemic examination was normal except for soft hepatomegaly. Fasting and random blood sugar values were

normal. Work up for tuberculosis and HIV infection was negative. Serum Complement C3 levels were markedly reduced. USG abdomen showed mild fatty liver. Urine albumin was repeatedly negative. In view of the gradual onset loss of subcutaneous fat from the face and upper trunk with relative sparing of lower part of body with low serum complement, a diagnosis of acquired partial lipodystrophy/ Barraquer Simons Syndrome (BSS) was made.

BSS, also known as acquired partial lipodystrophy, usually develops in the childhood<sup>1</sup> with onset being mostly before 15 years of age.<sup>2</sup> The typical pattern of fat loss seen in BSS is a gradually symmetric subcutaneous fat loss starting in the face and upper half of the body progressing downward with sparing of the lower abdomen and legs.<sup>3</sup> It is known to be associated with

**Figure 1a-b:** (a) Loss of buccal pad of fat over right cheek. (b) Normal appearing buccal pad of fat when the child was 5 years of age



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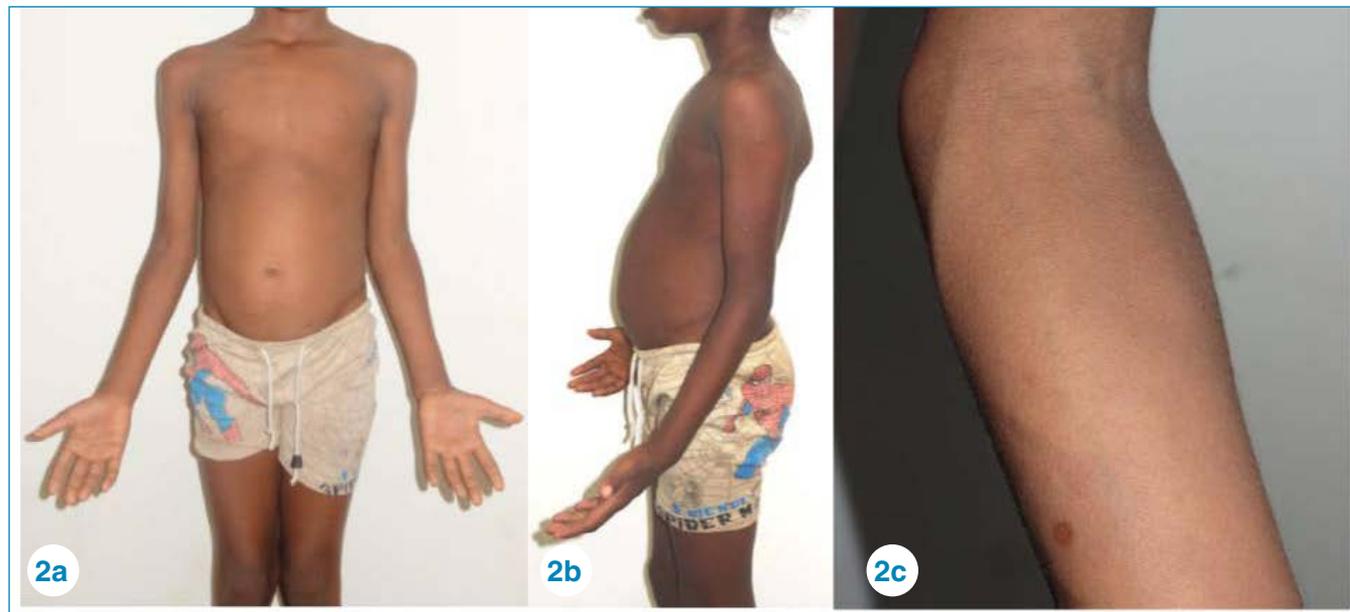
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**Figure 2a-c:** (a) frontal view (b) left lateral view of the child showing the fat loss pattern- more over the upper half of the body i.e the face, neck and upper limbs with relative sparing of the lower limbs. (c) Closer image of the upper limb showing the prominent loss of subcutaneous fat.



various autoimmune and metabolic disorders, most frequently with hypocomplementemia, and rarely with membranoproliferative glomerulonephritis.<sup>2</sup> The pathogenesis is believed to be due to autoimmune-mediated destruction of adipocytes.<sup>2</sup> Although many candidate genes have been described, the reason behind involvement of specific areas of fat loss remains an enigma.<sup>1</sup> The diagnosis of BSS is made on the basis of physical examination and hypocomplementemia. The prognosis of BSS is mainly dependent on renal involvement. A few patients have progressed to end-stage renal disease related to glomerulonephritis.<sup>4</sup> Affected children must be carefully followed up for renal involvement. Knowledge of this rare disorder

is useful during evaluation of a child with chronic wasting.

### References

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## Percutaneous Repair for Secondary Mitral Regurgitation: No Better Than Medical Therapy Alone

In a French multicenter study, 304 adults (mean age, 70; 75% men) with severe, symptomatic secondary MR (regurgitant volume >30 mL or effective orifice area >20 mm<sup>2</sup>) and an LVEF of 15% to 40% (mean, 33%) who were not candidates for surgery were randomized to percutaneous MitraClip repair plus medical therapy or medical therapy alone. MitraClip is used frequently and increasingly for secondary MR outside the U.S.

Technical device success was 96% of patients with attempted implantation. The primary composite endpoint and its components did not differ significantly between the intervention and control groups:

As per-protocol analysis of 246 patients produced similar results. Complications, including stroke, acute renal injury, and severe hemorrhage, were higher in the intervention group. Although repair was associated with a high and statistically significant reduction in MR grade (at hospital discharge, ≤2+, 92%; ≤1+, 76%), New York Heart Association class at 12 months was similar to that with medical therapy alone

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