

# Treatment of Dermatomyositis with JAK- inhibitor Baricitinib

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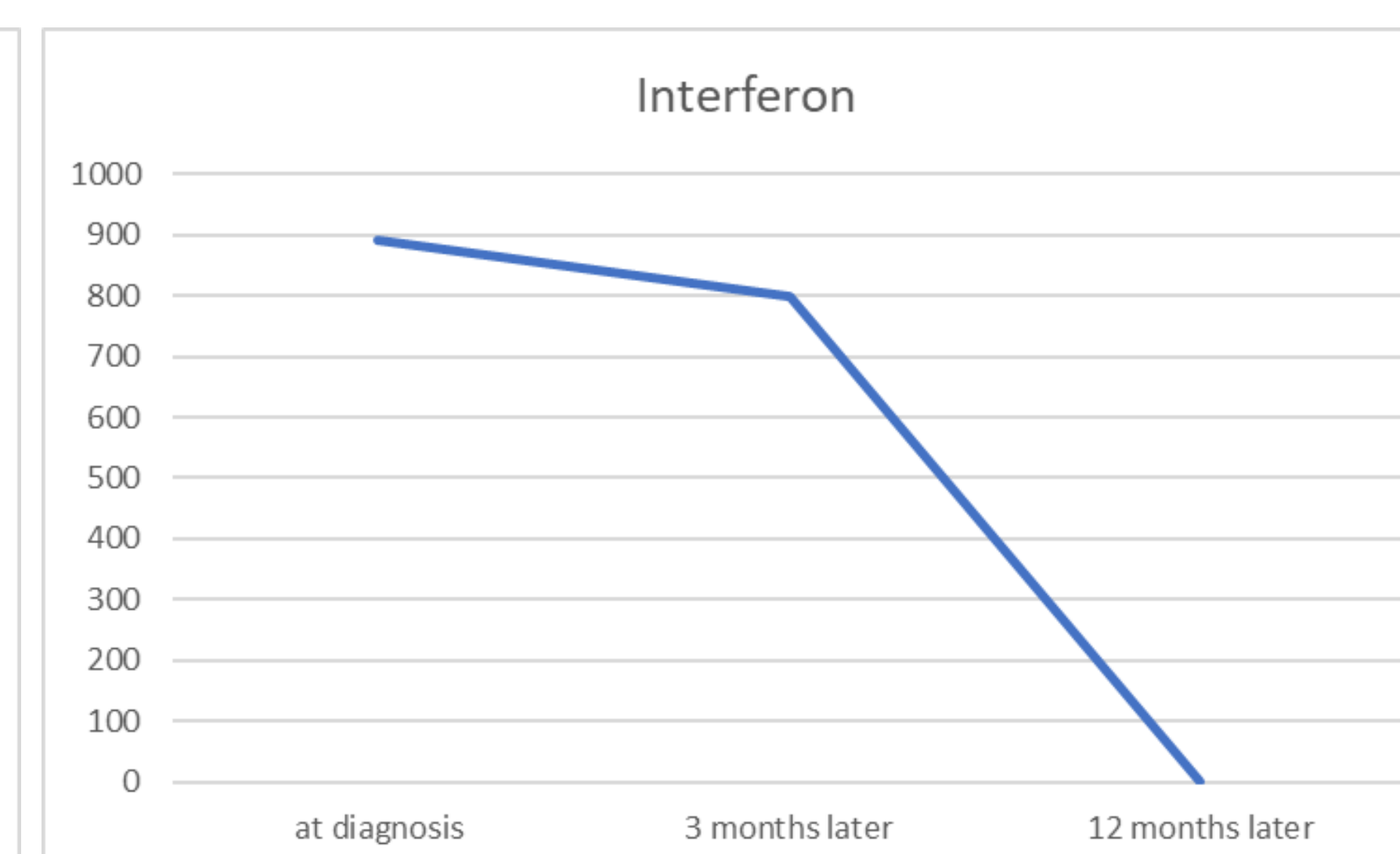
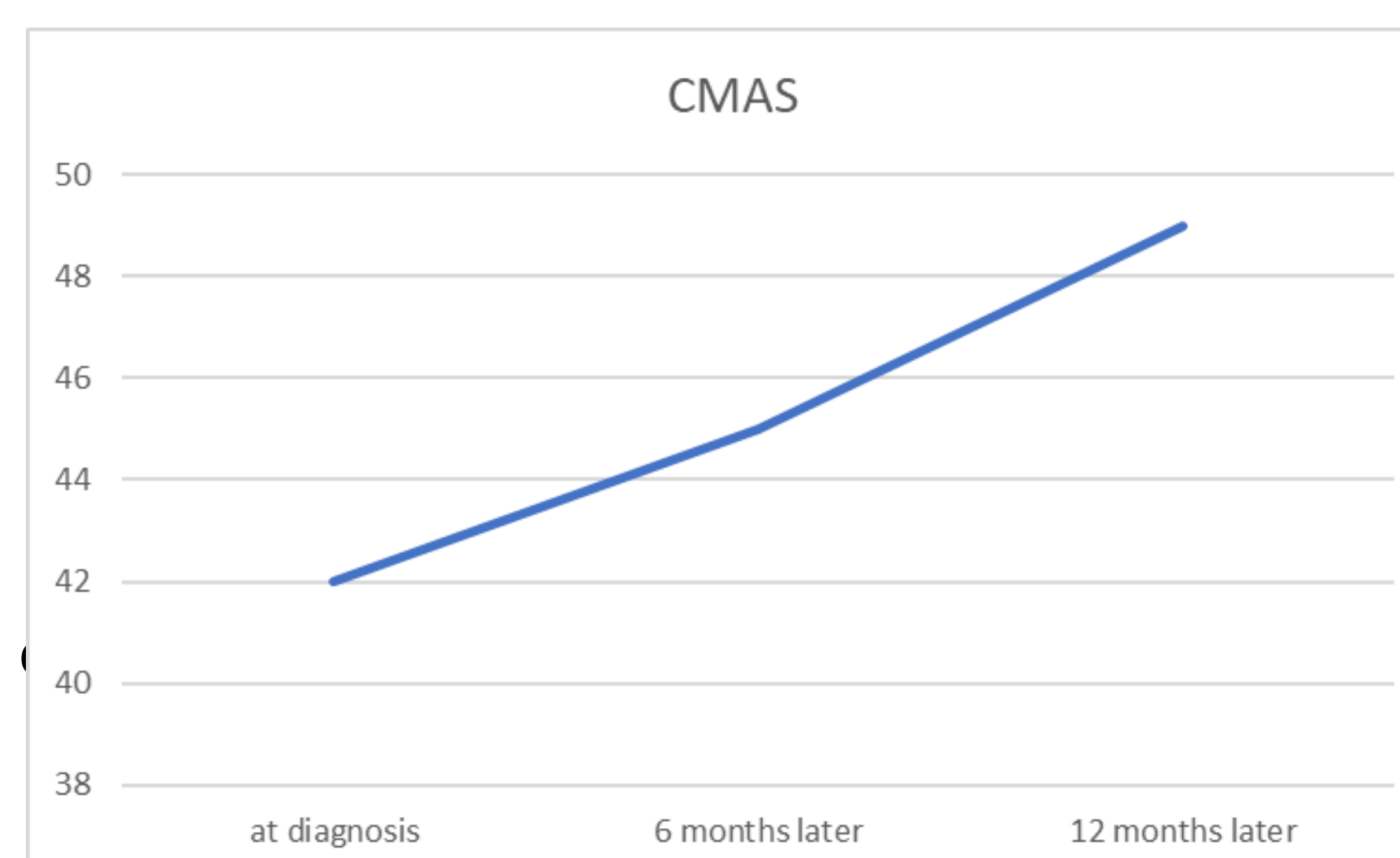
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## Background

Juvenile dermatomyositis (jDM) is a rare, idiopathic autoimmune disorder characterized by inflammation of both muscle and skin, often leading to significant morbidity and, in severe cases, mortality. Transcriptomic analyses in adult and juvenile forms of dermatomyositis (DM and jDM) have consistently demonstrated the upregulation of interferon (IFN)-regulated genes, suggesting a central role for the IFN pathway in the pathogenesis of the disease. Given the essential role of the JAK-STAT pathway in the pathophysiology of jDM, JAK inhibitors have emerged as a promising therapeutic option. Few case reports demonstrate the efficacy of JAK inhibitors in refractory jDM. Only a few patients treated with JAK inhibitors received Baricitinib.

## Case Report

We describe the successful use of JAK kinase inhibition in a 4-year-old boy with positive Mi2-alpha and MDA-5 antibodies, presenting with muscle weakness, arthritis, and skin involvement. The patient had crusted lesions on the hands, feet, nose, and ears, inversed Gottron's lesions, polyarthrititis, and muscle weakness. Laboratory tests showed Mi2-alpha and MDA-5 antibodies. MRI revealed widespread myositis, mainly in the gluteal region, along with synovitis and tenosynovitis in the elbows, hands, knees, and ankles. Inflammatory changes were noted in the skin, subcutaneous tissue, hepatosplenomegaly, and lymphadenopathy, but no pulmonary involvement or vasculitis. The patient had a highly elevated interferon signature (892.89, Ref. range: <12.49), prompting the exploration of treatments targeting the interferon pathway. In addition to standard therapies (IVIG, steroid pulse, hydroxychloroquine), Baricitinib (2mg/d) was introduced. The patient showed rapid improvement, with resolution of skin lesions, polyarthrititis, and normalization of muscle strength, without significant side effects.



CMAS:Childhood Myositis Assessment Scale

Interferon signature: ref. range: <12.49

## Conclusions

This case highlights the potential role of JAK kinase inhibition in managing patients with juvenile dermatomyositis and an elevated interferon signature, particularly in patients with MDA-5 antibodies. In our case, the use of JAK kinase inhibitor in the therapeutic regimen contributed to rapid resolution of symptoms and persistent disease control.

## References:

- Hinze, C.H., et al., *Development of practice and consensus-based strategies including a treat-to-target approach for the management of moderate and severe juvenile dermatomyositis in Germany and Austria*. *Pediatr Rheumatol Online J*, 2018. **16**(1): p. 40.
- Baechler, E.C., et al., *An interferon signature in the peripheral blood of dermatomyositis patients is associated with disease activity*. *Mol Med*, 2007. **13**(1-2): p. 59-68.
- Strauss, T., et al., *Rapid and sustained response to JAK inhibition in a child with severe MDA5 + juvenile dermatomyositis*. *Pediatr Rheumatol Online J*, 2023. **21**(1): p. 104.
- Sabbagh, S., et al., *Treatment of anti-MDA5 autoantibody-positive juvenile dermatomyositis using tofacitinib*. *Brain*, 2019. **142**(11): p. e59.
- Sener, S., et al., *Treatment with Janus kinase inhibitors in juvenile dermatomyositis: A review of the literature*. *Semin Arthritis Rheum*, 2024. **66**: p. 152426.

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