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A 36-year-old pregnant woman with hyperimmunoglobulinemia E

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Hyper-IgE syndrome (HIES) is an extremely rare multisystem disorder characterized by eczematoid dermatitis, recurrent staphylococcal cutaneous infections, recurrent pulmonary infections and elevated serum levels of IgE [1–2].

Characteristic facial appearance typically emerges in late childhood or early adolescence, including asymmetry, increased interalar distance, prominent forehead, deep-set eyes, broad nasal bridge, rough facial skin, prognathism, and high-arched palate [3]. In the course of the disease musculoskeletal, vascular and ocular abnormalities may also occur. The first signs of the disease may appear already in the newborn, but overall, the symptoms occur in late childhood or even early adolescence [1–3].

The treatment is prevention of abscesses, pneumonias with antibiotics, and aggressive treatment of infections, and sometimes using surgical procedures during abscess development.

A 36-year-old woman gravida III para II at 34 weeks and 4 days gestational weeks with a BMI 27 kg/m^2 (weight 70 kg; height 160 cm) was admitted to the hospital with symptoms of increasing respiratory failure and oligohydramniosis. For two days earlier she started to be dyspneic with a productive cough. In addition, she suffered from gastroesophageal reflux disease, recurrent dermatitis as well as infection of the upper and lower respiratory tract, multiple pathological rib fractures, a chronic obstructive pulmonary disease (COPD) with bronchiectasis. Her current pregnancy course was otherwise uncomplicated.

On admission, the patient had mild tachypnoea (20/min in sitting position); temperature was 36.6°C, blood pressure and heart rate were in normal ranges. Oxygen saturation was at a level of 89% and raised up to 97% while on L/min 100% oxygen with nasal cannula was administered. On chest auscultation reduced air entry were noted in the left lower lung, wheezes and dry rale. The SARS-CoV-2 antigen test was negative. The chest X-ray showed clinical and radiological signs of pneumonia.

After admission she received the first dose of betamethasone for fetal lung maturation and magnesium sulfate for fetal neuroprotection.

On the second day a decision was made to proceed with an urgent cesarean delivery due to further deterioration (Tab. 1). The boy infant was born prematurely in good general condition (Apgar scores 8/9/9; body weight 2150 g) and required only routine management after delivery.

On day three her oxygen saturations continued to decrease to 80% and 89% on 7 L/min of 100% oxygen. The chest high-resolution chest tomography revealed a tree-in-bud sign. The patient was treated with vancomycin 1 g twice per day and meropenem 0.5 g three times per day.

On seventh day, the C-section wound showed serous discharge and wound dehiscence. The bacterial swabs were taken and showed growth of methicillin sensitive coagulase-negative *Staphylococcus lugdunensis* susceptible to Vancomycin.

The patient fully recovered and was successfully discharged without any complications.

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Table 1. Results of the laboratory tests during hospitalization

	Admission day (0 day)	2nd day	4th day	6th day	9th day	12th day
CRP [< 5.0 mg/L]	8.5	6.2	72.3	170.0	69.9	5.49
PCT [≥ 2 ng/mL high risk of sepsis]	—	—	0.056	—	0.04	—
Hemoglobin [11.5–15.0 g/dL]	12.3	8.7	9.6	11.4	9.4	10.3
White blood cells × 10^3/μL [4–10]	11.6	12.24	11.64	13.11	10.79	8.91

CRP — C-reactive protein; PCT — procalcytonine

Eventually clinical improvement was achieved with vancomycin 1 g twice a day and meropenem 0.5 g three times a day intravenously. Long term prophylactic with cotrimoxazole may be taken into consideration (Tab. 1) [4]. As for neonate, the initial test showed no abnormalities.

On the fifth day postdelivery, an erythematous-papular rash appeared on the newborn's skin of the forehead and cheeks. Eventually the neonate was not diagnosed with Job's syndrome. Nevertheless, if such changes are found in a newborn of a parent suffering from Job's syndrome, specimens should be taken and subjected to histopathological verification. If typical histopathological changes are found, the newborn should be qualified for further diagnostics, despite the initially found normal IgE level [1–3].

The supervision of pregnancy of a patient with diagnosed Job's syndrome should include regular surveillance to detect signs of infection, serial ultrasound examinations, especially from 28th week with a careful assessment of fetal growth. Assessment of patient's pulmonary functions and dental inspection to prevent pulmonary complication and oral infection should be performed in each trimester [5].

Management of Job's syndrome should include careful control, protection of skin lesions with proper hygiene using moisturizing and antiseptic agents, and, if necessary, corticosteroid and antibiotic therapy.

Children, whose parents are living with HIES should be provided until adolescence with frequent, regular cardiological, neurological and angiological care as soon as possible.

Conflict of interest

All authors declare no conflict of interest.

REFERENCES

1. Hafsi W, Yarrarapu SNS. Job Syndrome. https://www.ncbi.nlm.nih.gov/books/NBK525947/ (05.01.2021).
2. Woellner C, Gertz EM, Schäffer AA, et al. Mutations in STAT3 and diagnostic guidelines for hyper-IgE syndrome. J Allergy Clin Immunol. 2010; 125(2): 424–432.e8, doi: 10.1016/j.jaci.2009.10.059, indexed in Pubmed: 20159255.
3. Cruz9Portelles A, Estopiñan A, ttp D. A new case of Job's syndrome at the clinic: A diagnostic challenge. Revista Portuguesa de Pneumologia (English Edition). 2014; 20(2): 107–110, doi: 10.1016/j.rppnen.2014.03.006.
4. Yousafzai, M. A., Nasim, K., Nizami, S. Q. (2000). Job's syndrome. Journal of Pakistan Medical Association, 50(12), 425-426. Available at: https://ecommons.aku.edu/pakistan_fhs_mc_women_childhealth_paediatr/542 (05.01.2021).
5. Lindenbaum C, Chatwani A, Oyer R, et al. Hyperimmunoglobulinemia E and pregnancy: a case report. Am J Obstet Gynecol. 1987; 157(5): 1273–1274, doi: 10.1016/s0002-9378(87)80312-5, indexed in Pubmed: 3688089.