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FRI0569

WHAT DOES IT MEAN TO BECOME PREGNANT WITH JUVENILE IDIOPATHIC ARTHRITIS? A MONOCENTRIC EXPERIENCE IN A TERTIARY CENTRE OF MILAN DEDICATED TO YOUNG ADULTS AFFECTED BY JIA

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Background: During the last seventeen years, biological and non-biological therapies have been used in our center in an open prospective study for the treatment of refractory JIA not only in paediatric age but also in young adults. The availability of more effective drugs for JIA has dramatically increased the number of patients willing to become pregnant. Data regarding the effect of JIA on pregnancy outcome are scant.

Objectives: The study aims to evaluate the capability of JIA patients to become pregnant, the effects of drugs on pregnancy outcome and the impact of gestation on the disease course.

Methods: JIA patients regularly followed up in our Transition Clinic of the Division of Rheumatology, Gaetano Pini Institute of Milan, were enrolled in the study at positive pregnancy test. During the whole pregnancy patients underwent monthly clinical examination and obstetric ultrasound. Data regarding disease activity and pregnancy course (fetal morphometric parameters, fetal heart rate, fetal growth, Doppler velocimetry) were collected. Gestational age, birth weight and APGAR score were also recorded.

Results: 29 pregnancies in 23 women affected by refractory JIA became pregnant during a 17 years follow up. All patients had a long lasting polyarticular disease (median duration of 23 years) not responsive to DMARDs, and became pregnant during biologic therapy. Patients were treated with Etanercept (11 patients), Golimumab (4 patients), Rituximab (3 patients), Adalimumab and Certolizumab (2 patients respectively) as monotherapy, and in most of the cases after multiple switches. One woman was treated with Etanercept during the first pregnancy and Adalimumab during her second pregnancy. Three patients decided for an elective termination and 3 experienced an early miscarriage; among 23 pregnancies resulting in live born infants, only 3 had premature births and 1 cleft palate. A pregnancy was complicated by gestosis, two by placental detachment. All the babies were followed up during the 17 years of observation and did not experience any major late complication. Eight patients were subjected to intra-articular infiltrations during pregnancy to switch off the disease, 18 patients resumed therapy shortly after childbirth and only 7 patients decided to breastfeed.

Conclusion: despite a large amount of studies demonstrating the safety of anti-TNF during pregnancy, data regarding the effects of biologics on pregnancy outcome in JIA are still lacking. The very low number of patients treated with traditional DMARDs achieving low disease activity underline the pivotal role of new biologic drugs in the management of aggressive long standing JIA, to improve the quality of life of these patients, including family planning. In our experience, no greater number of unexpected complications or side effects were observed in JIA patients during pregnancy compared to the other autoimmune diseases. Discontinuation of therapy has increased the risk of flare, requiring local therapy, confirming that EULAR recommendations for the use of biologics during pregnancy can be applied also in JIA.

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CLINICAL CHARACTERISTICS IN PATIENTS WITH PEDIATRIC SYSTEMIC LUPUS ERYTHEMATOSUSDIAGNOSED BEFORE 6 -YEARS OLD IN A THIRD-LEVEL HOSPITAL IN MEXICO

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Background: Pediatric-onset Systemic Erythematosus Lupus (pSLE) is known to be more aggressive than adult-onset SLE. The disease is extremely rare under the age of 5 years and with severe course.

Another big difference is the association of pSLE with monogenic primary immune deficiencies (PID). Some PIDs have been associated with SLE or lupus-like manifestations: deficiencies of the classical complement pathway, female carriers of X-linked chronic granulomatous disease, IgA deficiency (present in 5% of pSLE).

It is important to consider PIDs in patients with severe manifestations in pSLE, in our country there are no studies of this association.

Objectives: To describe the clinical manifestations, laboratory features, characteristics and therapeutics of patients with diagnosis of SLE before 6 years of age at National Institute of Pediatrics in Mexico City

Methods: This is a retrospective study with the review of the patient's records with diagnosis of SLE before 6 years old, from the Immunology department between January 2005 and December 2016 at National Institute of Pediatrics in Mexico City.

Results: Between 2005-2016 there were 295 newly patients diagnosed with pediatric Systemic Lupus Erythematosus (pSLE) at our hospital, from these patients 2.7% (8) were diagnosed with pSLE before 6 years of age. According to this, 7 (87.5%) were female. The youngest patient was 26 months at moment of diagnosis.

Every patient had at least 4 criteria of the ACR classification for SLE. Only 2 (25%) had malar rash at diagnosis, and 1 (12.5%) presented discoid rash. Seventy-five percent of patients presented glomerulonephritis at diagnosis and also 75% had an Hematological disorder.

In terms of serology, 37.5% had positive native DNA antibody, 37.5% antibody to Sm protein and 37.5% antiphospholipid antibodies. One hundred percent of patients had presence of antinuclear antibody (ANA) at moment of diagnosis.

Conclusion: During 2005-2016 we found 295 newly patients with diagnosis of pediatric Systemic Erythematosus Lupus (pSLE), from these, eight patients (2.7%) were diagnosed before 6 years of age. Seventy-five percent presented recurrent infections and 50% with severe infections, and one patient died due to septic shock.

Systemic erythematosus lupus with an early onset has a more severe phenotype and has been associated with monogenic primary immune deficiency, presenting with severe infections and higher mortality.

In our country, we do not have genetic testing to help us determine the endless monogenic defects associated with early-onset SLE, being necessary to determine etiology and specific treatment as needed.

We recommended that in every patient with severe pSLE who develops life-threatening, infections or due to unusual germs, associated with hypocomplementemia and/or consanguinity, to perform an approach to rule out any primary immune deficiency.

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