ABSTRACT
Hereditary angioneurotic edema (HAO) is the clinical translation of C1 esterase inhibitor deficiency (C1 INH). The frequency and severity of clinical manifestation are extremely variable, ranging from edema of moderate extremes to obstruction of the upper airways. The natural history of HAO during the pregnancy is unpredictable. The hormonal impregnation, the trauma of the childbirth appear factors susceptible to activate the crises. We report a case of OAH and pregnancy. Through this case as well as a revue of the literature, we are going to try to identify the clinical manifestations of this rare affection during the pregnancy and in post-partum as well as the various modalities of therapeutic coverage.

KEYWORDS: Hereditary angioneurotic edema; C1 esterase deficiency; Pregnancy; Treatment.

INTRODUCTION
The hereditary angioneurotic edema (HAO) is a rare disease autosomique dominant, due to a deficit in inhibitor of esterase C1 (C1 INH) which regulates the activity of the first component of the complement[1]. Its prevalence is considered at 1/100 000 cases in France.[2] THE HAO is characterized by recurring crises of edema affecting the skin, the mucous membranes, the tract gastroenteritis - intestinal and respiratory tracts. The major risk is to develop an inflation of all the aerodigestif tract with a big risk of obstruction of the superior air traffics.[3] The studies concerning the OAH and the pregnancy are rare with few data on the relation between this affection and pregnancy.[3]

OBSERVATION
A.N 32 years old, primigravida, followed for angineurotique oedema diagnosed in pre-pubertal period further to the appearance of segmental and spontaneously resolutive edemas. The repetitive aspect of edemas, as well as the family context (the father and the sister are affected by the disease) incited to make a study of the complementary system. The dosage of C1 INH was collapsed confirming the diagnosis of hereditary angioneurotic edema (HOA) of type I by quantitative C1 INH deficiency. The patient was put on corticoids at the time of the crises with a good answer. She had a multidisciplinary follow-up (obstetricians and resuscitator-anaesthetists) since the diagnosis of her pregnancy. At the beginning of the pregnancy, the patient presented a threat of early abortion stopped by the taking progestagens with active minimal dose. The first crisis of edema (interesting the face) arose following this threat” figure 1” was quickly solved by taking corticoids by oral route (méthyl prednisolone in dose of 60 mg / j by oral route during 3 days following the crisis).

Figure 1: Crise d'œdème angioneurotique héréditaire se manifestant par un œdème facial.

During her follow-up, the prenatal assessment was abnormal; the patient presented light crises affecting extremities at the rate of a crisis a month. In 28 weeks of amenorrhea the patient was admitted in the service for management of a moderated threat of premature delivery associated with a moderate abdominal crisis confirmed in the abdominal ultrasound by the presence of low abundance ascite. The obstetric ultrasound was without particularity. The patient answered well to the injectable corticoids (méthylprednisolone 80 mg it intra venous during 3 days) and the threat of premature delivery was
stopped by the calcic inhibitors (the nicardipine in intravenous drip relayed by oral route).

The obstetric ultrasound of growth done in 32 weeks of amenorrhea showed a biometrics between 10 and 25 ème percentile without anomaly in the umbilical doppler. In 34 weeks of amenorrhea, the ultrasound control showed a harmonious intra uterine growth retardation with a biometrics lower than 3 ème percentile and a fetal weight estimate of 1400g, without oligoamnios, a zero umbilical diastole, a decrease of the resistance index at the cerebral Doppler with a negative wave on the ductus venosus “Figure 2”, where from the indication of an extraction by high way as a matter of urgency.

Figure 2: Negative wave on Doppler of ductus venosus.

The caesarian allowed the extraction of a male newborn child Apgar 10/10 the birth with a weight of 1400g. The newborn child was transferred in neonatal resuscitation where he stayed for breeding. No particular event arose during the caesarian (a demand/request) for the reserve of freshly frozen plasma seen the not availability of the concentrated of C1-INH, or during the hospitable(hospital) stay.

The patient left the service 5 days later and the newborn child left the neonatal resuscitation after three weeks with a weight of 2000 g. Three months after the childbirth, the mother and the child are in good shape.

DISCUSSION

The angioneurotique oedema hereditary is an autosomic disease Dominant produced by a deficiency of functioning of C1-INH. C1-INH is a plasmatique protease which works to surround the activation of the complement and the formation of the bradykinine on several sites in the sequence. The absence of this enzyme allows an activation freely of the stunt of the complement, what pulls a clinical angio-oedema.

In 1888, Osler describes the effects of OAH during the pregnancy by attributing the premature delivery and the rate of stillbirths to the process of the disease. Since then, some isolated reported cases, described the evolution of the OAH during the pregnancy. The majority of these reported cases suggested an increase of the frequency of the crises during the pregnancy as well as for the period of the post-partum. Our patient also has an increase of the frequency of the crises during the pregnancy but, she presented no crisis in the post-partum.

The fertility and the rate of abortions are identical to those

Found in the normal population

The typical treatment implied the preventive use of danazol (or other androgenic agents) in doses titled to prevent attacks. Antifibrinolitic agents, the tranexamic acid especially, are less effective than androgens, but tend to be used at the prepubescent child's or if androgens are besides badly tolerated or are against indicated.

Glucocorticoids are active in high doses (500 - 1000 mg of intravenous Méthylprednisolone). During the pregnancy, it is acceptable to continue treatment with tranexamic acid. Danazol is against indicated. The treatment of the severe crises is based on the use of concentrated C1INh. Our patient received only a symptomatic treatment by corticoids in low doses at the time of the crises with a good response. A case reported of symptomatic treatment of OAH during pregnancy requiring several hospitalizations at the time of the crises. Our patient benefited from two hospitalizations, one for abdominal crisis of OAH having caused a threat of premature delivery and the second for caesarian. There was no case of symptomatic treatment by corticoids nor the limitation of the fetal growth during the pregnancy, reported in the literature.

The management of delivery depends on the evolution of the pregnancy. If the patient presented grave episodes with increase of their frequency, the childbirth must be covered by the concentrated of C1 INH (20U/kg in drip intra venous). If the disease is less grave, the disease prevention by the concentrated of C1 INH is not necessary. However, it must be available at time of childbirth if need. In our case, the not availability of the concentrated of C1 INH, was replaced by a request for the reserve of freshly frozen plasma. The analgesia epidural is strongly recommended in case of OAH. The rate of caesarian is not higher at these patient's than in the general population. The indication of the caesarian in our observation was for severe limitation of the fetal growth.
CONCLUSION

THE OAH is a rare but potentially dangerous pathology which owes
Be known by the obstetricians and the anaesthetists to know well the processing of the crises but also the preventive treatment of the patients affected by OAH during the follow-up of the pregnancy, the perioperative care and in post-partum.

REFERENCES