

# TAKAYASU and pregnancy, a case report and review of the LITERATURE

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**Abstract:** *Takayasu disease is a primary arteritis of large and medium-sized vessels that primarily affects the aorta and its major dividing branches. Pregnancy in conjunction with Takayasu disease increases the risk of cardiovascular complications. We will present a case of a parturient followed for Takayasu disease and taken in charge in the maternity of the university hospital centre Hassan 2 in FÃ's with a favorable outcome of her delivery.*

## Introduction

Takayasu disease is a chronic inflammatory arterial disease of unknown etiology, primarily affecting the aorta and its branches. It is a predominantly female disease. In concomitant pregnancies with this disease or after diagnosis, the risk of cardiovascular complications, such as hypertension and congestive heart failure, is high [1]. Only a few cases have been described in the literature, hence the interest of our case.

## Medical observation

This is Mrs B.A, 39 years old, with a scarred uterus (obstetric scar), followed for TAKAYASU disease in internal medicine at the HASSAN II University Hospital of Fez for 9 years, with a moderate inflammatory syndrome in the biological check-up and a total occlusion of the middle portion of the right subclavian artery and a narrowing of the left axillary artery without frank stenosis in the angiography of the suprasuperior arteries. ECG, transthoracic ultrasound and brain scan came back without abnormalities.

She is a fourth gesture with a history of two vaginal deliveries and a caesarean section in the context of pre-eclampsia giving birth to two boys and a girl of unclear birth weight, currently aged 17 years, 15 years and 7 years, all with good psychomotor development.

She was put on acetylsalicylic acid; long term oral corticosteroid therapy (60 mg prednisone/d) and methotrexate stopped before conception.

The patient was referred to us for follow-up of her pregnancy at 5 months (1st consultation) contracted on the pill.

Antenatal monitoring revealed balanced blood pressure figures on diet alone. We assessed fetal well-being by ultrasound monitoring which showed normal growth; normal fetal Doppler and by outpatient cardiac rhythms which returned normal. The pre-eclampsia work-up was normal.

The patient was followed up in prenatal consultations at a rate of one consultation per week with monitoring of blood pressure figures measured in the lower limbs, as well as a pre-eclampsia check-up, a fetal heart rate and a doppler ultrasound of the uterine and umbilical arteries (which were normal). It should be noted that she did not have any flare-ups of her condition during the entire pregnancy.

She was admitted to hospital at 32 weeks' gestation for closer monitoring and a decision was made on the delivery route depending on the progress.

She gave birth at term, by vaginal delivery, to a newborn male, Apgar: 10/10, birth weight: 3300g. The postpartum period was simple, without complications.

## Discussion

Takayasu disease is an arteritis of large and medium-sized vessels, the management of which is complicated by the absence of effective diagnostic criteria, validated therapeutic strategies and reliable activity criteria. Indeed, it is a pathology that can manifest itself by cardiovascular, neurological, ophthalmological... etc. Microscopic examination shows a segmental panarteritis with a predominantly medial-adventitial appearance. The diagnosis is based on a combination of clinical history, physical examination,

clinical suspicion and vascular imaging techniques. The aetiology is unknown and treatment is aimed at controlling the inflammatory process and preventing secondary sequelae, particularly systemic hypertension [2]

Pregnancy can be revealing of the disease (especially revealing or aggravating pulmonary arterial hypertension), which is the case in a quarter of pregnancies occurring during the course of Takayasu disease [3, 4]. It is reported to have an adverse effect on the course of the disease in 61% of cases [5]. Cardiovascular complications seem to be more frequent and are to be feared in the perinatal period (strokes and cardiac decompensation during the third trimester and perinatally [5, 6]). Maternal mortality is estimated at 5% according to Wong who reported two cases of death, one of which occurred on the fourteenth day postpartum due to myocardial infarction [6]. Therefore, patients with Takayasu disease should be monitored very closely before and during pregnancy for strict blood pressure control.

Fetal complications in Takayasu disease are those of hypertension. Fetal hypotrophy has been observed in 18% of cases by Ishikawa [7]. It is due to hypertension and damage to the hypogastric vessels, which can lead to utero-placental hypoperfusion [8]. Wong has proposed a score to assess fetal prognosis [6] (Table 1) based on the existence of abdominal aorta and renal artery involvement, third trimester blood pressure, date of onset of pre-eclampsia and onset of correct management [8]. (TABLE I: Prognostic score for neonates of mothers with Takayasu) Each parameter is scored from 0 to 2: if the score is below 4, the fetal prognosis is favorable. All newborns with a score of 4 or more had a weight below the 3rd percentile. In our case, we had a good prognostic score of 1.

In order to improve the maternal-fetal prognosis, it is advisable to plan the pregnancy by first establishing a baseline vascular lesion assessment, and to allow the pregnancy only if the possible complications are prevented. [8]. In some cases, contraception will have to be started for a period of time, in particular during treatment with an immunosuppressant. In other cases, pregnancy will be possible while continuing treatment, under regular medical supervision [3].

The management of the pregnancy must be bi-disciplinary, cardiological and obstetrical, with regular monitoring of blood pressure, which can be difficult to measure if the humeral arteries are affected. Biological monitoring will include urea, creatinine, proteinuria, transaminases and platelets [1, 3].

Fetal status is assessed by ultrasound to estimate fetal growth and doppler, and by fetal heart rates. The authors recommend preventive treatment with anti-platelet agents based on acetyl salicylic acid at a dose of 100 mg / in case of hypertension or valve prosthesis [5, 8, 9].

During pregnancy, corticosteroid therapy may be instituted or continued. Hospitalization of the patient may be justified as early as 32 weeks' gestation for better maternal-fetal monitoring [8], which was the case for our patient.

In the event of a relapse of the disease during pregnancy, the recommended treatment usually combines boluses of Solu-Medrol® 15 mg/kg per day (not exceeding 1 g) for three consecutive days with prednisone 1 mg/kg per day. In refractory cases, azathioprine may be introduced [3]. Hypertension should be treated aggressively with calcium channel blockers or alpha and beta blockers.

Delayed treatment results in an unfavorable environment for the fetus. Ishikawa and Matsuura [7], and Wong et al [6] reported a high incidence of intrauterine growth retardation and hypertension, and involvement of the maternal abdominal aorta has been cited as an etiological factor for the same.

Pre-eclampsia has been noted by several authors [3, 6]. Indeed, the blood pressure level determines the course during pregnancy [3, 10]. Wong reported 11 cases out of 15 full-term pregnancies [6]. However, blood pressure is difficult to measure when the vascular involvement is brachial and hypertension is only diagnosed when measured in the lower limbs [4, 5, 6]. In addition, pregnancies reported in Takayasu disease are at high risk of hypertension and eclampsia, particularly when there is renal artery involvement. The pulmonary arteries are involved in almost half of Takayasu disease cases [3].

Vaginal delivery is allowed with perfect lesion control. Prophylactic caesarean section is reserved for severe arteriopathy with poorly controlled hypertension, arterial aneurysm or heart failure [4, 8]. Our patient gave birth vaginally. Epidural analgesia is recommended as it reduces the hypertensive peaks caused by pain. Instrumental extraction is desirable to shorten expulsive efforts [8].

In the postpartum period, the use of oxytocin is permitted but ergot derivatives are contraindicated. Breastfeeding is not contraindicated [5]. Prevention of thromboembolic disease is necessary [8].

Estrogen-progestin contraception is contraindicated. The prescription of new minidosage progestogen molecules is possible in young non-hypertensive women with a mild form of the disease and no other risk factors. Mechanical contraception using an intrauterine device is possible in the absence of contraindications, particularly valvulopathy or anticoagulant treatment [8].

## Conclusion

Pregnancy is permissible in cases of uncomplicated Takayasu disease. However, it can be fraught with serious complications threatening the vital prognosis of the mother (pre-eclampsia, cardiac decompensation) and the fetus (intra-uterine growth retardation); hence the need for planning and cardio-obstetrical monitoring of the pregnancy.

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TABLE I: Prognostic score for newborns of mothers with Takayasu

Score	Abdominal aortic involvement	Start of management	Median blood pressure in 3rd trimester (mm Hg)	Pre-eclampsia
0	No	1st trimester	> 100	No
1	Yes	2nd trimester	101-130	3rd trimester
2	Yes + renal artery involvement	3rd trimester	< 130	1st ou 2end trimester