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Gynecology

Pregnancy Due to Drepanocytosis with Maternal and Fetal Complications: Clinical Case and Focus

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Abstract **Clinical Case**

The aim was to describe the monitoring and management of maternal and fetal complications of this composite SC sickle cell pregnancy. Pregnancy is a high risk situation for a woman with sickle cell disease and for her fetus. Management must be multidisciplinary and involve obstetricians, doctors specializing in sickle cell disease, hematologists, organ specialists, anesthetists, resuscitators, midwives. Sickle cell disease can make pregnancy worse, but pregnancy can also make the disease worse. Ocular involvement of sickle cell anemia is common, particularly in SC and Sβ+-thalassemic sickle cell patients. The severity of ophthalmologic complications is mainly linked to these proliferative retinal manifestations. Laser treatment is used to control the growth of new blood vessels in the eyes affected by this disease. We report the case of a major sickle cell pregnancy with fortuitous discovery of maternal retinopathy and intrauterine growth retardation in the fetus. From this case we will describe the problems that can cause this category of pregnancy and make an update on the management.

Key words: SC major sickle cell disease, pregnancy, complications.

ABBREVIATIONS
CNRST: National Center for Scientific and Technological Research: Sickle cell disease CRLD: research and control center
CPN: prenatal consultation
FMOS: Faculty of Medicine and Odontostomatology
GS/ Rh: Blood group: Rhesus
HbF: fetal hemoglobin
HbS: Hemoglobin S
HbC: hemoglobin C
Immunoglobulin G: IgG
Immunoglobulin M: IgM
Rhesus: Rh
RPR: Rapid plasma reagin
TPHA: Treponema Pallidum Hemagglutination Assay)
VDRL: Venereal Disease Research Laboratory.

INTRODUCTION

Sickle cell disease is one of the genetic disorders of hemoglobin in which the abnormal hemoglobin S is found in high concentrations in red blood cells. It is an autosomal recessive disease of variable expression. This is a substitution of a adenine by a thymine (GAGpGTG) resulting in the replacement, at the level of the β globin chain, of glutamic acid in position 6 by a hydrophobic valine [1, 3]. There are 3 main major sickle cell syndromes (SS, SC, Sß thalassemia).

Sickle cell disease is a worldwide disease. However, populations of African origin are particularly affected. In some West African countries, 30 to 40% of

the population is believed to carry the gene. In France, the Paris region and the French overseas departments and territories represent the two main areas of predilection where sickle cell disease is rife [3].

Despite the improvement in life expectancy and quality of life thanks to the diagnosis and management of sickle cell syndromes during pregnancy, this is a high-risk situation for women with sickle cell disease and for their fetus [3, 4]. In developing countries, which cannot benefit from this early treatment, the rate of maternal and fetal morbidity and mortality is still very high. Management must be multidisciplinary and involve obstetricians, doctors specializing in sickle cell disease, hematologists, organ specialists, anesthetists, resuscitators, midwives. Sickle cell disease can make pregnancy worse, but pregnancy can also make the disease worse [3, 4, 5].

Sickle cell disease increases the risk of developing complications in pregnancy and conversely, pregnancy promotes the development of sickle cell complications. Maternal mortality still remains at 1% despite improved care. Serious maternal complications are high blood pressure and pre-eclampsia, thromboembolic events and infections (very common urinary tract infections). The fetal risk is significant: intrauterine growth retardation, hypotrophy, fetal death in utero, prematurity, etc. During pregnancy, acute complications of sickle cell disease are likely to occur with greater frequency, in particular Vaso-occlusive complications (CVO), chest pain syndrome (ATS), urinary tract infections and pyelonephritis, aggravation of anemia. In the latter case, the repercussions are both fetal and maternal even in patients who previously had little symptom [7, 8, 11].

Ocular involvement of sickle cell anemia is common, particularly in SC and Sβ + -thalassemic sickle cell patients. Ocular involvement is estimated to affect approximately 15-20% of adult patients with homozygous sickle cell (SS) and 35-40% of adults with SC. Eye involvement mainly affects the retina. It is a peripheral vascular occlusion which constitutes the main element of sickle cell retinopathy and which is the cause of complications: neovascularization, intravitreal hemorrhages, retinal detachment. Involvement of the macula (about 30% of retinopathy cases) puts you at risk of blindness. The severity of ophthalmologic complications is mainly linked to these proliferative retinal manifestations. Laser treatment is used to control the growth of new blood vessels in the eyes affected by this disease [8, 10, 11]. Our objective was to describe the problems generated by this category of patient and to focus on the management.

PATIENT AND METHOD (Presentation of the Case)

Mrs. K; D.D. 30 years old, third gesture second par, two living children (G3P2V2) was known SC sickle cell disease and monitored at the Center for Research and the Fight against Sickle Cell Disease in Bamako Mali. Her husband's electrophoresis was not available at the time of our treatment. Mrs. K; D.D. lived in bakorobabougou, a suburb of the district of Bamako. The patient came to our outpatient clinic on her own to monitor her pregnancy. Previously, she had performed a first obstetric ultrasound which showed an evolving mono-embryonic intrauterine pregnancy of ... ... The initial examination noted in her medical history a composite SC sickle cell anemia (electrophoresis of hemoglobin seen). The general examination found a fairly good general condition, blood pressure at. 110 / 65mmhg and Weight 58kg. On physical examination, the conjunctivae were moderately colored, the uterine height was 24 cm, the mobile cephalic presentation and the sounds of the fetal heart perceived with the Pinard stethoscope were at 1150 btt / min. When touched vaginally, the cervix was long, posterior, soft and closed. We had concluded at the diagnosis of active pregnancy of 28 weeks. The updated Hb electrophoresis returned to SC comprising the different fractions: HbF = 7.9%, HbS = 49.0%, HbC = 43.1%. The remainder of the follow-up laboratory workup ANC gave the following result: GS / Rh = O positive, Blood count: Hb = 10.3g / dl, Hematocrit = 29.9%, red blood cells = 3.02.106 / mm3, white blood cells = 10.1.103 / mm3, VGM = 99.3µm3, platelet count = 102000 / mm3, glycemia = 3.97mmol / L, azotemia = 3.83mmol / l, search for albumin and sugar in the urine was negative, toxoplasmosis serology (IgG = negative, IgM = negative), rubella serology IgG = positive, IgM = positive), syphilis serology with TPHA + VDRL = negative, detection of antigen against viral hepatitis B (AgHVB) was negative. We had implemented monthly monitoring up to 28NT then every 15 days until 36NT. An opinion from the hematologist of the CRLD was requested and the latter to monitor the disease throughout the pregnancy. During an appointment with the CRLD doctor, a routine eye examination by the ophthalmologist requested concluded with the diagnosis of proliferative retinopathy of the left eye. The management of this retinopathy by the ophthalmologist was done by laser photocoagulation. The ophthalmologist then recommended delivery by scheduled cesarean section, the vaginal delivery of which could worsen serious retinal damage and would compromise the patient's vision during pushing efforts during childbirth. Thus at 36 weeks of pregnancy monitoring the cesarean section was performed on / 11/11/2021.

During pregnancy, the prevention of anemia was ensured by the administration of folic acid and tardyferon B9 (only for proven iron deficiency (ferritinemia) and screening for urinary tract infection was provided by monthly ECBU and at the slightest suspicion of urinary tract infection, cefixime was prescribed.

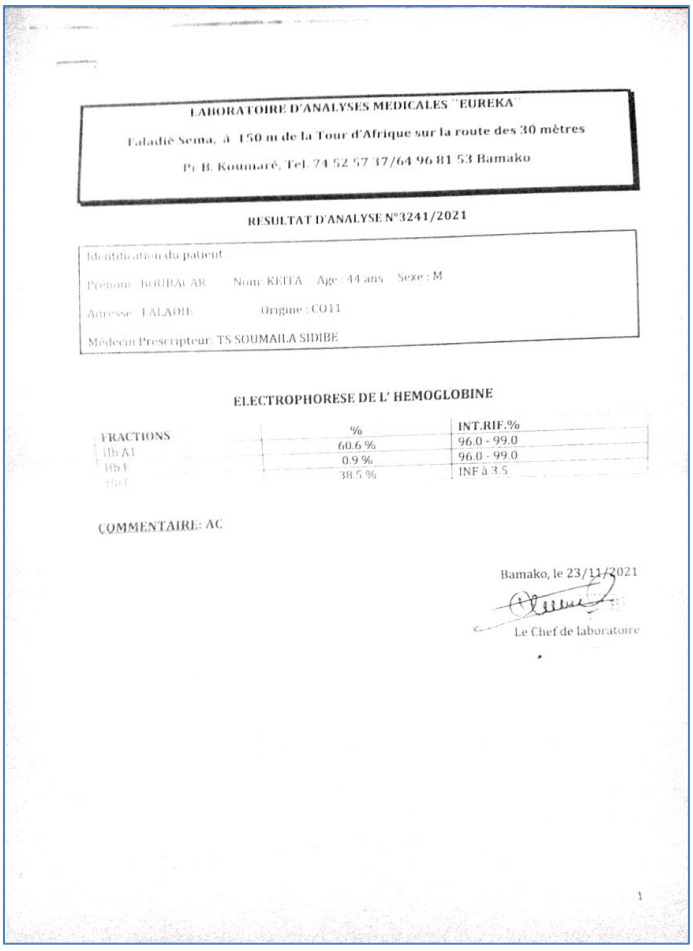
The delivery was done by caesarean section as recommended by the ophthalmologist, the advantage of which was to protect the retina from this which could be damaged during relapses at the time of expulsion. The low transverse segmental cesarean section allowed cephalic extraction of a hypotrophic newborn with a birth weight equal to 2260g, height = 49cm and head circumference = 32cm, Apgar score = 9/10 at the 1st minute and 10 / 10 in the 5th minute. The newborn was immediately invited to the pediatrician for neonatology. The type of anesthesia was spinal anesthesia with oxygenation and there was no intraoperative blood transfusion. Breastfeeding was the method of feeding the infant mother.

In post-partum the postoperative treatment in the first 24 hours included hydration due to 3l / 24 hours, painkillers: Paracetamol infusion and injectable nefopan every 6 hours in case of pain, prevention of maternal infection (endometritis, urinary tract infection) by wearing clean packing and prescribing clamoxyl 2G / day / 07days and at the end early active mobilization. We did not have to prescribe a blood thinner for this case. The postoperative follow-up was favorable and the patient and her newborn baby were allowed to be discharged from the hospital on the 3rd day of her operation with dressing appointments every three days. Until healing and an appointment for the gynecological medical examination. Months after the operation.

DISCUSSION

The limitations of our study included, among other things, the inaccessibility of examinations such as maternal and fetal Doppler ultrasound, prenatal diagnosis, genetic counseling, etc. All pregnant women from Africa and the West Indies should be tested for hemoglobinopathy by hemoglobin electrophoresis at the first antenatal visit. If they are carriers of the sickle cell trait, electrophoresis of the spouse should be performed. If both spouses are carriers of the trait, genetic counseling should be offered [3].

This recommendation was part of our daily practices since, according to national guidelines (Mali), the request for hemoglobin electrophoresis should be part of the mandatory examinations of the biological assessment of the first consultation. In our case, we had counseled the couple on the interest of performing electrophoresis of the spouse as well as the two living children. What had been done and the results were as follows: Spouse, B K the profile was AC with the fractions HbA1 = 60.6%, HbF = 0.9% and HC = 38.5%.



LABORATOIRE D'ANALYSES MEDICALES "EUREKA"
Faladiè Sema, à 150 m de la Tour d'Afrique sur la route des 30 mètres
Pr B. Koumaré, Tel: 74 52 57 47/64 96 81 53 Bamako

RESULTAT D'ANALYSE N°3241/2021

Identification du patient :
Prénom : BOUBACAR Nom : KEITA Age : 44 ans Sexe : M
Adresse : FALADIE Origine : CO11
Médecin Prescripteur: TS SOUMAILA SIDIBE

ELECTROPHORESE DE L' HEMOGLOBINE

FRACTIONS	%	INT.RIF.%
HbA1	60.6 %	96.0 - 99.0
HbF	0.9 %	96.0 - 99.0
HbC	38.5 %	INF à 3.5

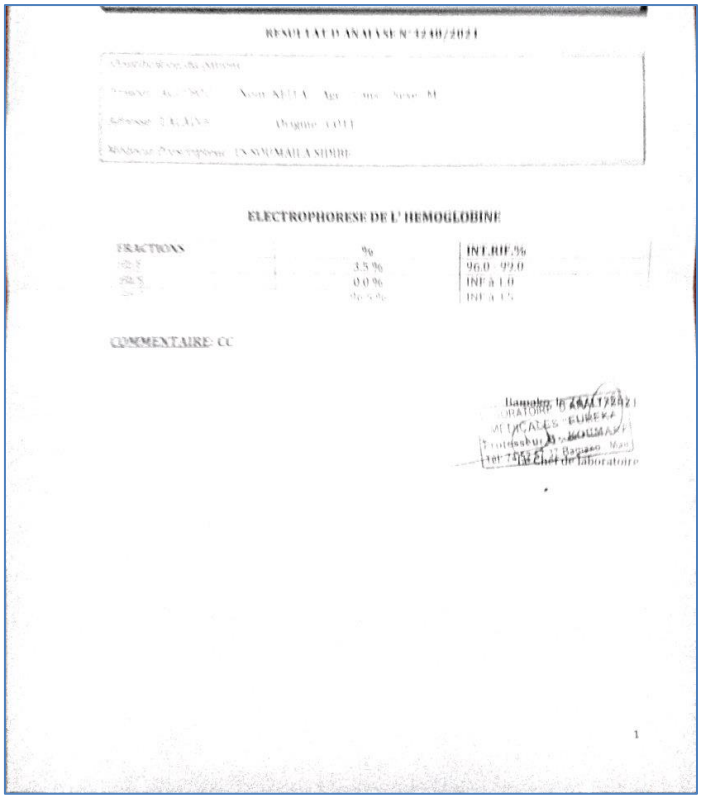
COMMENTAIRE: AC

Bamako, le 23/11/2021

Le Chef de laboratoire

1

Boy 1, AK, the profile was CC with the fractions HbF = 3.5%, HbS = 00% and hemoglobin C = 96.5%



RESULTAT D'ANALYSE N° 1240/2021

ELECTROPHORESE DE L' HEMOGLOBINE

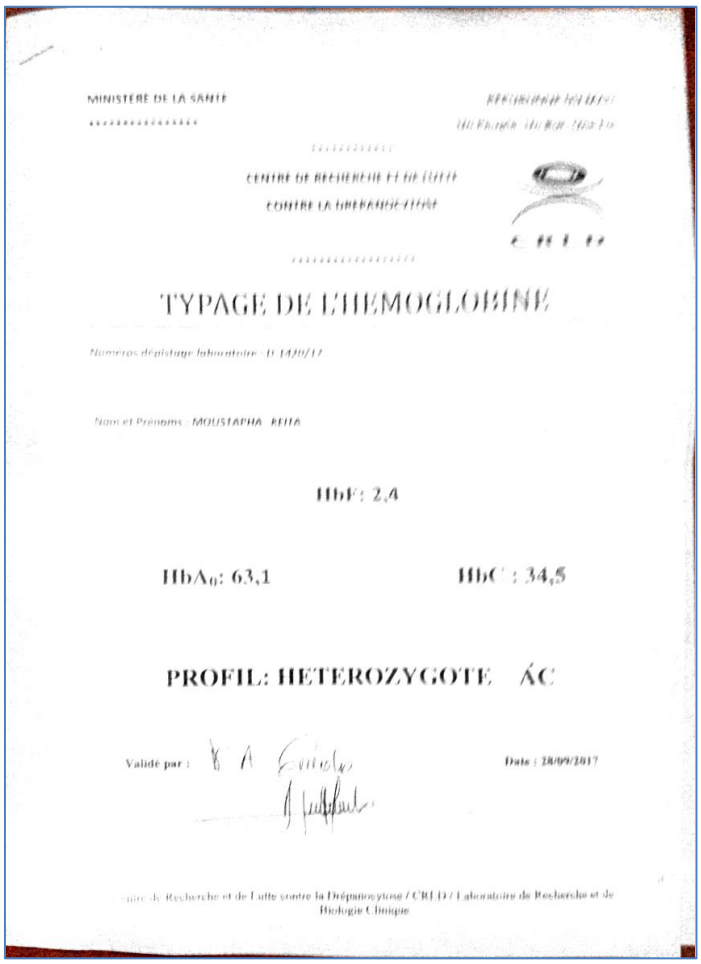
FRACTIONS	%	INTERF.%
	35 %	96.0 - 92.0
	0.0 %	INF à 1.0
		INF à 1.5

COMMENTAIRE: CC

Le Chef de laboratoire

1

Boy2: M K, the profile was heterozygous AC with the fractions were HbF = 2.4%, HbA0 = 63.1% and HbC = 34.5%



MINISTERE DE LA SANTE

CENTRE DE RECHERCHE ET DE LUTTE CONTRE LA DREPANOCYTOSE

TYPAGE DE L'HEMOGLOBINE

Numéros dépistage laboratoire : D 1420/17

Nom et Prénoms : MOUSTAPHA KEITA

HbF: 2,4

HbA_0: 63,1

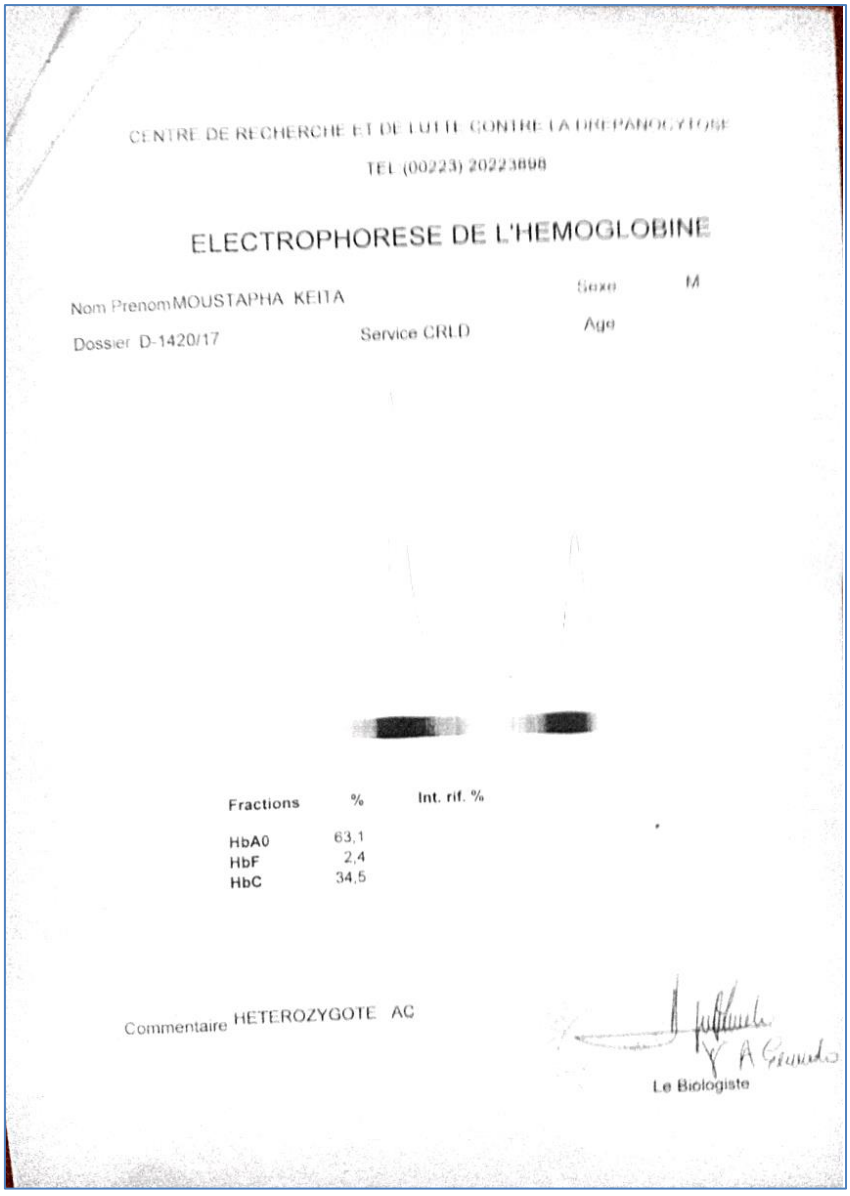
HbC : 34,5

PROFIL: HETEROZYGOTE AC

Validé par :

Date : 28/09/2017

Centre de Recherche et de Lutte contre la Drépanocytose / CRELD / Laboratoire de Recherche et de Biologie Clinique



CENTRE DE RECHERCHE ET DE LUTTE CONTRE LA DREPANOCYTOSE

TEL (00223) 20223898

ELECTROPHORESE DE L'HEMOGLOBINE

Nom Prenom MOUSTAPHA KEITA Sexe M

Dossier D-1420/17 Service CRLD Age

Fractions	%	Int. rif. %
HbA0	63,1	
HbF	2,4	
HbC	34,5	

Commentaire HETEROZYGOTE AC

Le Biologiste

Regarding genetic counseling, we did not have until now at the risk of misleading national jurisdiction, which would be one of our limitations in our case study.

The pregnancy should be monitored by an obstetrician accustomed to this pathology in association with the referring physician for hemoglobinopathies. In our case, this recommendation was respected, allowing the diagnosis of the ophthalmic complication (proliferative retinopathy [3, 5]).

Assessment of the general condition during visits: (anemia, haemolysis, liver and kidney functions, etc.) [3,5]. Our management complied with this recommendation since we had prescribed folic acid due to 1compx2 / day and tardyferon B9 as well as the monthly realization of the ECBU in order to prevent severe complications such as severe anemia and urinary tract infections that can compromise the outcome of this pregnancy. Before and during the cesarean operation at the end of the post-perfume period, we had not performed a transfusion in this patient because the monitoring had enabled us to maintain a Hb level above the values indicating a transfusion as well as her clinical condition of this patient. In the literature opinions are divided on transfusion and it does not make sense to transfuse systematically, each case will have to be managed on a personalized basis. Routine transfusion did not improve the obstetric prognosis [3, 5.10].

The delivery in our case was done by cesarean section in order to avoid the development of complications that could impair or cause loss of sight caused by the excessive growth of blood vessels in the back part of the eye [10, 11]. Clarisse Bonkian in a study on sickle cell anemia and pregnancy had obtained 50% of cesarean section [10].

In the postpartum period, hydration and the prevention of infections, especially endometritis, were

provided according to the recommendations in the literature [3, 5, 10, 11]. For our part, we had not prescribed an anticoagulant but we had recommended early emergence and active mobilization in the immediate postpartum period. Counseling for a contraceptive method had escaped our notice, but it was scheduled for the next medical visit a month after the operation.

CONCLUSION

Pregnancy in sickle cell anemia patient is a high risk pregnancy. The multidisciplinary follow-up allowed the detection and effective management of retinopathy by laser photocoagulation during pregnancy. Until then, it has allowed this patient to keep her vision even if a fetal hypotrophy that may be related to this pathology was observed.

Conflict of interest

The authors declared no conflict of interest

CONTRIBUTIONS

The study design was made by Bourama Kané, Seydou Mariko and S Sanata The methodology was adopted by P Coulibaly and S Mariko. Editing and analysis was carried out by S Mariko and S Sanata. B Kané, A Traoré MB Coulibaly NS Bagayogo M Haïdara S Khalil A Sidibé K Tembiné M Traoré A Saye had read and approved the final document before its submission

Thanks

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