DOI: 10.21276/sjmcr.2016.4.11.18

Scholars Journal of Medical Case Reports **ISSN 2347-6559 (Online)**
Sch J Med Case Rep 2016; 4(11):869-873 **ISSN 2347-9507 (Print)**

(An International Publisher for Academic and Scientific Resources)

Pediatric extracorporeal membrane oxygenation in homozygous sickle cell patient for acute chest syndrome.

Papa Salmane Ba[1], Lionel Berthomieu[3], Marie-Pierre Castex[4], Bertrand Leobon[2], Fabio Cuttone[2]
[1]Department of Cardiovascular -Thoracic Surgery, Fann University Hospital, Dakar, Senegal.
[2]Department of Cardiac Surgery, University Hospital (CHU), Toulouse, France.
[3]Department of Pediatric Intensive Care Unit, University Hospital (CHU), Toulouse, France.
[4]Department of Hematolgy and Oncology Pediatric Unit, University Hospital (CHU), Toulouse, France.

***Corresponding author**
Papa Salmane Ba, MD
Email: salouba@gmail.com

Abstract: Extracorporeal membrane oxygenation is used in Sickle cell disease patients is unclear. We present a case of a 6 years old girl with homozygous hemoglobin SS disease complicated with chest syndrome. Oxygenation was maintained with two days of VA extracorporeal membrane oxygenation and 30 days of VV ECMO.
Keywords: Sickle cell disease, hemoglobin SS disease, oxygenation.

INTRODUCTION

The term sickle cell crisis is principally a clinical term embracing a number of pathological mechanisms including vaso-occlusion, bone marrow aplasia, and red cell sequestration. In homozygous sickle cell anemia (Hb SS) vaso-occlusive crises are the most common cause of morbidity and mortality in children and adults. Acute chest syndrome (ACT) remains a life-threatening complication of sickle cell disease (SCD). It concerns half of patients and results in 25% of deaths and account for more than 90% of hospital admission [1, 2].

Extracorporeal membrane oxygenation (ECMO), in particular, venoarterial (VA) ECMO is used routinely in the treatment of severe respiratory and cardiovascular failure in neonatal, pediatric and adult patients. However, the role for ECMO in Sickle cell disease patients is unclear [3].

We present a case of sickle chest syndrome in which oxygenation was maintained with extracorporeal membrane oxygenation, which allowed the pulmonary changes to resolve.

CASE REPORT

A 6 years old girl with homozygous hemoglobin SS disease (SCD) was admitted in a context of acute thoracic pain. Suspicion of acute right pneumonia was made and did not evolve well with Ceftriaxone. In the worsening of respiratory state with extension of the acute thoracic syndrome on the whole right lung complicated by anemia, it was admitted at the intensive care unit. At admission, she was non-invasive ventilated with 6 cm H_20 of positive end expiratory pressure. Haemoglobin was 5,6g/dl and Haemoglobin S was 70%.

Erythrocytapharesis and blood transfusion allowed an increase in haemoglobin (8,8g/dl) and decrease in haemoglobin S (33%).

Subsequently, she presented an acute respiratory distress with a refractory hypoxia (Sao_2 86%) despite a F_IO_2 at 100% and hemodynamic instability, 12 hours after admission. Lactate rate was 6,8mmol/L. She was intubated and mechanical ventilation was initiated. Chest radiograph demonstrated bilateral patchy pulmonary opacities (figure 1). After an initial clinical improvement, she developed a cardiovascular failure requiring a high dose of inotropic and ECMO support in femoro femoral configuration to maintain hemodynamic status.

Cannulation of right subclavian artery was unsuccessful because of small diameter of vessels. A 12 Fr cannula was surgically placed in the right common femoral artery. A reperfusion whith a 6 Fr sheath was connected to the arterial line. The right common femoral vein was cannulated with a 19 Fr cannula.

ECMO assistance was initiated with optimal flow, varying between 1.5 l/min to 1.9 l/min. A 2D transthoracic echocardiography demonstrated a severe biventricular impairment of contractility. Anticoagulation with heparin was initiated with a target activated clotting time (ACT) of 130s-188s.

Cardiac function recovery was good after two days of assistance. She was able to sustain her systemic perfusion on epinephrine 0,1µg/kg/min and

norepinephrine 0,4µg/kg/min. The patient was gently weaned off ECMO to 0,9 l/min flow with a normal biventricular function normal lactates state.

In the worsening of chest X-ray (figure 2) and blood gas (Po_2 72mmHg and Pco_2 83mmHg on 100% F_IO_2), a switch into VV ECMO with an Avalon Elite® Bi-Caval Dual Lumen Cannula n°19 in the right jugular vein under 2D transthoracic echocardiographic guidance (figure 3) was decided. VV ECMO allowed recovering of lung compliance, blood exchange without organ and tissue dysfunction (figure 4). Recruitment maneuvers and corticosteroid therapy with 2 mg/kg/Day begun in Day 10 and a bolus of 500mg/m2 at day 25; improved respiratory status (figure 5).

She was weaned from VV ECMO after 30 days in front of a satisfactory respiratory recovery confirmed by radiologic findings (figure 6) and the good respiratory tests (Po_2 154 mmHg, Pco_2 53 mmHg, Sao 98% with flow pump rate of 1,5l/min on 35% F_{IO2} (table 1).

After repeated packed red blood cell transfusion, haemoglobin was 10g/dl and haemoglobin S lower than 20%.

Brain monitoring made with non-invasive cerebral oxygenation (NIRS Covidien®), transcranial Doppler and electroencephalography didn't show any complication.

Mechanical ventilation was stopped after one week; she was discharged at home 2 months after her admission. After 12 months on Follow-up, she has a good clinical state, without respiratory insufficiency and normal neurological outcome.

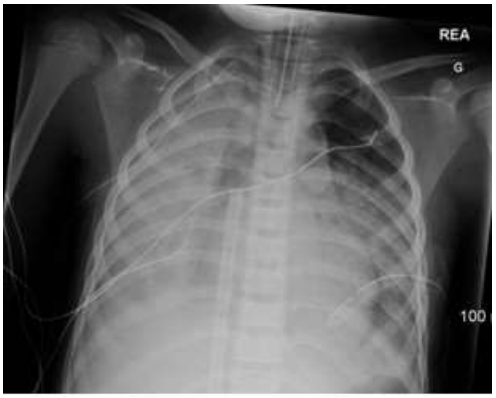

Fig-1: Va (Veino arterial) Ecmo Day 2

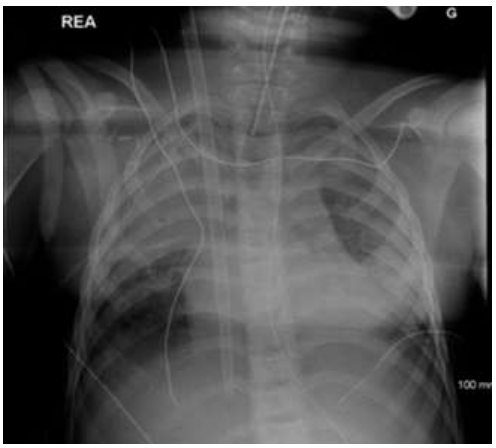

Fig-2: Vv (Venovenous) Ecmo Day 5

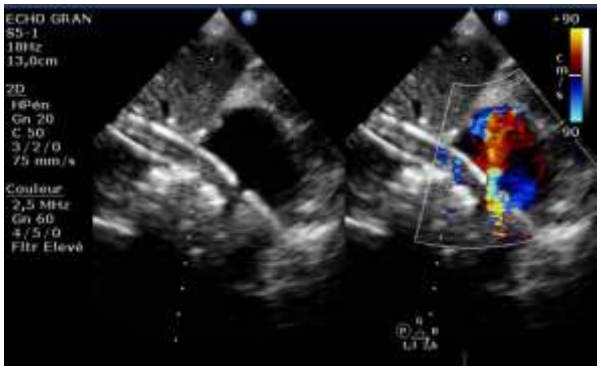

Fig-3: Avalon Elite® Bi-Caval Dual Lumen Cannula n°19 in the right jugular (red arrow)

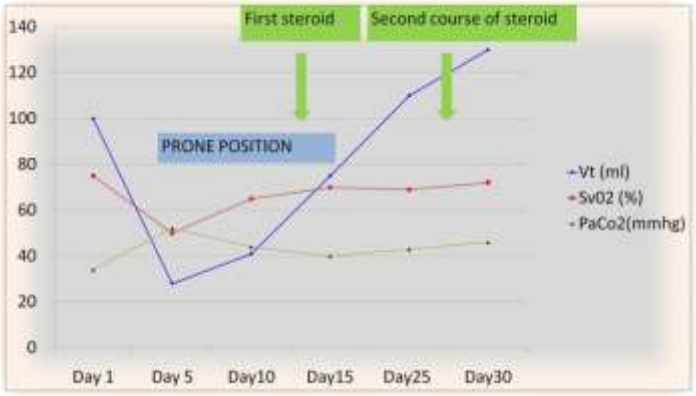

Fig-4: Evolution and combines treatments

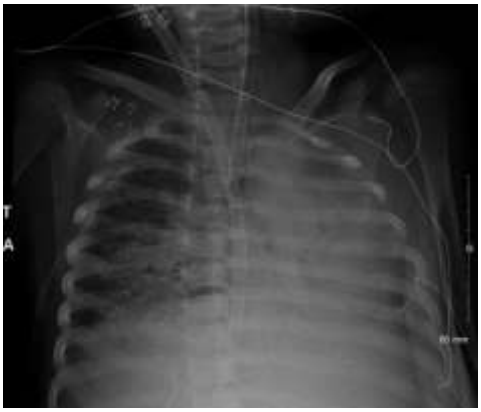

Fig-5: Vv Ecmo Day 20

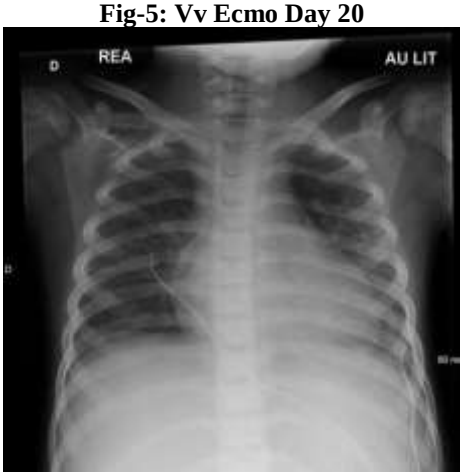

Fig-6: Day 40(Vv Ecmo weaned)

Table-1: Sao2 Monitoring with Fio2 every two hours in day 30 Vv Ecmo

(Sao_2 : %)	94	96	96	96	96	99	99	99	98	98	98	99	99	98	98
(F_Io_2 :%)	75	100	100	80	70	60	55	50	55	55	55	55	40	40	35

DISCUSSION

Vaso-occlusive crises are a common and potentially fatal complication of sickle cell disease. They are most common in homozygous sickle cell anemia and may be recurrent [4]**.** Acute pulmonary disease is a common and serious consequence of sickle cell disease [5]. The increased viscosity inherent in deoxygenated deformed sickle cells and oxygenated, irreversibly sickled cells is the major factor responsible for obstruction of blood flow in the microvasculature [6]. In addition, the elongated rigid sickle erythrocyte is less deformable and flows through capillaries with difficulty. A vicious cycle is set up whereby the above factors promote stasis which, in turn, augments local hypoxia and acidosis and leads to increased sickling and ultimately to microvascular occlusion and ischaemic infarction [7]**.** In the chest syndrome, it would thus appear that extensive ischaemic necrosis can occur in sickle cell anaemia without morphological evidence of thrombosis of small or large vessels.

Seventy-five per cent of patients who develop an acute chest syndrome have been admitted with a painful crisis. Our patient has an [Pao_2/F_Io_2] =80mmHg before ECMO therapy. The mainstay of any acute sickle cell crisis including the acute chest syndrome is simple or exchange transfusion to reduce the HbS fraction (aiming for <30%). ECMO support is considered when a patient remains in hypotensive shock despite maximal treatment and the clinical condition is believed to be potentially reversible [8]**.** It is the case of our patient.

We preferred to swith in VV-ECMO two days after VA-ECMO because she has a good heamodynamic state contrasting with a bad respiratory state (Po_2 72mmHg and Pco_2 83mmHg on 100% F_Io_2). Another explication is the fact that the registry data seem to suggest that those undergoing VV vs. VA ECMO have lower mortality and less neurological morbidities [3].

The monitoring of VV ECMO is quite difficult and it is based on the arteriovenous oxygen difference more than arterial Po_2 that remains low during assistance. The use of Po_2 level alone can mislead treatment decisions.

We preferred internal jugular cannulation strategy for VV-ECMO to avoid the risk of recirculation of oxygenated blood, poor lower leg venous drainage and difficulties in patient mobilization in femorofemoral connection [9]. The use of heparin remains controversial as there is also some evidence that heparin is beneficial in vaso occlusive crises [9]. Patient was anticoagulated with unfractionated heparin, using target activated clotting time of 130s-188s. Use of corticosteroid doesn't decrease prognosis if needed [10], perhaps the level of Hb S under 20% reduces the risk as before surgical procedure for patient with sickle cell disease. Hour patient was going well after introduction of corticosteroid therapy. We kept the Hb S level under 20% to minimize thrombosis and hemolysis risk.

Our patient has a good evolution with clinical, functional, and radiological improvement. These results demonstrate a promising application of ECMO support in acute chest syndrome associated with a corticosteroid treatment.

CONCLUSION

Our experience with this patient highlights both the potential benefits and associated risks of VA ECMO and VV ECMO. Without ECMO, our patient wouldn't survive. It's why we will do a cohort study to confirm that this method of treatment should be considered in the management of severe sickle chest syndrome when standard methods of artificial ventilation fail.

REFERENCES

1. Anionwu E, Walford D, Brozović M, Kirkwood B. Sickle-cell disease in a British urban community. Br Med J (Clin Res Ed). 1981 Jan 24;282(6260):283-6.
2. Athanasou NA, Hatton C, McGee JO, Weatherall DJ. Vascular occlusion and infarction in sickle cell crisis and the sickle chest syndrome. Journal of clinical pathology. 1985 Jun 1;38(6):659-64.
3. Kuo KW, Cornell TT, Shanley TP, Odetola FO, Annich GM. The use of extracorporeal membrane oxygenation in pediatric patients with sickle cell disease. Perfusion. 2013 Sep 1;28(5):424-32.
4. Diggs LW. Sickle cell crises: Ward Burdick award contribution. American Journal of Clinical Pathology. 1965 Jul 1;44(1):1-9.
5. Davies SC HPScd. Sickle cell disease. Br J Hosp Med, 1984; 31(6):440-4.
6. Chien S, Usami S, Bertles JF. Abnormal rheology of oxygenated blood in sickle cell anemia. Journal of Clinical Investigation. 1970 Apr;49(4):623.

7. Klug PP, Lessin LS, Radice P. Rheological aspects of sickle cell disease. Archives of internal medicine. 1974 Apr 1;133(4):577-90.
8. Nevet A, Polak T, Dagan O, Waisman Y. Extracorporeal Membrane Oxygenation as a Resuscitation Measure in the Pediatric Emergency Department. The Israel Medical Association journal: IMAJ. 2015 Oct;17(10):639-41.
9. Parhar K, Parizkova B, Jones N, Valchanov K, Fowles JA, Besser M, Telfer P, Kaya B, Vuylsteke A, Rubino A. Extracorporeal membrane oxygenation for the treatment of adult sickle cell acute chest syndrome. Perfusion. 2015 Jun 30:0267659115593172.
10. Ogunlesi F, Heeney MM, Koumbourlis AC. Systemic corticosteroids in acute chest syndrome: friend or foe?. Paediatric respiratory reviews. 2014 Mar 31;15(1):24-7.