The role of antifibrinolytics in sickle cell disease during open heart surgery in pediatrics
(An open discussion for a case of TOF scheduled for total correction)

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A 2.5 years old male patient (10 Kg) with sickle cell disease (about 50% sickling on Hb electrophoresis), with ToF scheduled for total repair. Medical and surgical history were irrelevant.

Clinical examination:
HR = 100 /min with sinus rhythm
BP=100/70
Chest: clear
Rectal temperature: 37 degree Celsius
SO2% on room air was 78%

Laboratory investigations:
CBC: Hb= 15 gm/dl, Pcv =43 , Plats = 187  10³/ml , WBC = 11  10³/ml, Pt =14 sec.
Apart from slight increase in liver enzymes, liver and Kidney function tests were within the normal.

Chest X-Ray:
showed( BOOT SHAPED HEART); increased cardiothoracic ratio .

ECG:
Right ventricular hypertrophy

Echocardiography:
Large Mal-alignment of VSD with less than 50% overriding of aorta.
Infundibular and Valvuler PS with maximum pressure across RVOT=75 /mmHg.
Reasonable size MPA (RPA: 9mm LPA: 8mm) Mc/Goon ratio:1.9.
Good LV size & contractility.

Anesthetic management:
The patient was premedicated with l.M. ketamine.
G.A.was induced with, i.v fentanyl and sevoflurane mask in 100% oxygen.
Mechanical ventilation with 100% oxygen to maintain PCO2 at 30-35 mmHg.
Anesthesia was maintained with isoflurane and fentanyl(10-30mcg/kg)as needed to maintain systemic blood pressure and SPO2% .

CPB was initiated using a membrane oxygenator (liliput II) , roller pump and proper tubing set. Priming was done using both colloids (200 ml FFP) & crystalloids (100 ml Ringer lactate solution) and 100 ml packed red blood cells. Heparinization was achieved with 30 mg heparin (3 mg/kg)
targeting ACT > 400 sec. CPB was conducted using aortobicaval cannulation. After aortic cross-clamping, cardiac arrest was done by antegrade blood cardioplegia 300 ml (30 ml/kg) repeated every 25 min. During CPB we avoided active cooling of the patient by just drifting temperature to about 32 c.

On bypass HCT was maintained with 28%. & ABG was maintained to avoid acidosis (PH=7.35 – 7.44). Haemodynamics were maintained with MBP=45-50mmHg with pump flow = 120 ml/kg/min. VSD closure and transanular patch were done.

Towards the end of CPB, patient’s blood was hemofiltered to raise the HCT to 33%. Aortic cross-clamp time estimated was 70 min. Weaning of CPB was accomplished uneventfully.

Adrenaline 100 nanogram/kg/ min. and dobutrex 10 µg/kg/min were used as inotropic support. Protamine (60 mg) to neutralize heparin effect was given. ACT showed 180 sec. ABG post bypass was done, HCT: 32%. One unit of PRBCS was transfused. Hemostasis was started, generalized oozing was noticed which was unsuccessfully controlled in 30 mins. 2 units of FFP were transfused. Extra i.v. protamine 20mg and cyclocapron (100mg/kg) were given. Finally successful control of bleeding was achieved. Closure of the chest was achieved and the patient was transferred to ICU. Post-operative recovery was smooth and he was discharged home in good condition.

**Question to be discussed:**

1. WHAT IS THE ROLE OF EXCHANGE TRANSFUSION BEFORE CARDIAC SURGERY OR DURING CPB?
2. PHARMACOLOGICAL AND NON-PHARMACOLOGICAL METHODS TO DECREASE BLEEDING.
3. PERIOPERATIVE OPTIMIZATION OF THE SICKLE PATIENT’S CONDITION AS REGARDS HB, HTC, PLTS, PT, PTT, INR.
4. WHAT KIND OF BLOOD OR BLOOD PRODUCTS TO BE INFUSED.