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Limiting Blood Transfusion in Sickle Cell Disease –A Single Centre Study

KEYWORDS: Sickle cell anemia, Blood transfusion

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ABSTRACT

Introduction: Transfusion with the blood of normal hemoglobin (AA) has the potential to relieve the symptoms and complications of SS disease. Conditions like splenic sequestration, acute chest syndrome and aplastic crisis can be fatal if not treated with blood transfusion. Present study was aimed to assess the impact of protocol limiting blood transfusion in sickle cell disease patients. Study design-Retrospective study conducted in a cohort of sickle cell patients. Methodology-Study was conducted in sickle cell centre of Government Medical College and hospital, Akola. Patients selected were registered patients at sickle cell centre. Patients were divided into two groups-first group before Jan 2011 (before implementation of protocol to limit blood transfusion in sickle cell patients) and second group after the implementation of protocol (Feb 2011 to March 2015). Result-Out of total 80 registered patients 47 (59%) were male and remaining 33 (41%) were females. Before protocol average per person-year blood transfusion was 1.55 blood transfusions/Person/Year. Similar way after implementation of protocol average per person-year blood transfusion was 0.37 blood transfusions /Person/Year. This shows blood transfusion requirement was reduced by approx 75% of previous transfusions. Rate of blood transfusion was 4.18 times more before uniform protocol implementation. Conclusion- Large numbers of sickle cell patients are treated in central India at peripheral centres and private practitioners by blood transfusion for all complaints. There is a need to create awareness among all clinicians about indications of transfusion and hazards of multiple transfusions in these patients.

Introduction-

Sickle cell disease is an autosomal recessive genetically transmitted haemoglobinopathy responsible for considerable morbidity and mortality. It is prevalent in many parts of India including central India, where the prevalence in different communities has ranged from 9.4% -22%[1,.2]. Considerable clinical diversity has seen in India in patients with sickle cell anemia.[3,4].

Transfusion with the blood of normal hemoglobin (AA) has the potential to relieve the symptoms and complications of SS disease. Conditions like splenic sequestration, acute chest syndrome and aplastic crisis can be fatal if not treated with blood transfusion [5, 6]. In patients with sickle cell disease (SCD) blood can be administered as a simple top up transfusion or as an exchange transfusion where there is simultaneous removal and replacement of blood. Splenic sequestration is most common in early childhood. There is massive blood pooling in the spleen causing severe anemia and hypovolemia and its immediate treatment is correction of hypovolemia with RBC transfusion.

Acute chest syndrome describes a new pulmonary infiltrate with respiratory finding with cough, dyspnea or new onset hypoxia in a patient with SCD [7]. Management includes respiratory therapy, antibiotics and RBC transfusion. Transfusion improves oxygenation within 12 to 24 hours of RBC administration. Perioperative conditions including suboptimal hydration, poor oxygenation and academia can lead to SCD related complications such as acute chest syndrome, painful vaso-occlusive episodes and infections. Perioperative simple transfusion to achieve hemoglobin of 10g/dl is effective in preventing postoperative complications [8]. Chronic severe anemia from kidney failure may need treatment with periodic blood transfusions. Chronic transfusion programme for prevention of stroke is the accepted indication [9].

Complications of blood transfusion-the main side effects of blood transfusion in patients with SCD are alloimmunisation [10], hyperhemolytic transfusion reactions,iron overload [11] and transfusion transmitted infections.

Alloimmunization rates are reported to run between 18 and 36% in patients with SCD who have received transfusion and rates increase as the number of units given rises. Red cell antibodies to Rh and Kell antigen systems are most important [12]. Patients with multiple antibodies can be virtually untransfusable and it can be difficult to obtain blood in an emergency.Red cell antibodies can become undetectable over time and further exposure to the relevant antigens can cause an anamestic response and delayed hemolytic transfusion reactions(DHTR) which can have severe clinical sequel [10,13]. Recommended treatment is transfusion avoidance and immunosuppression with intravenous immunoglobulin, corticosteroids, erythropoietin and rituximab [13,14,15].

Risk of iron overload is there with frequent blood transfusions [11]. Very high levels of iron can lead to hemosiderosis, which can be fatal if untreated. Transfusion acquired

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infections like HIV and hepatitis C have shown a marked decrease but still present a major risk. Patients who are iron overloaded are particularly vulnerable to *Yersinia enterocolitica*. With time, venous access becomes a potential problem which makes it impossible to continue transfusions [16].

Methods-

The present retrospective study was conducted in Government Medical College Akola situated in western Vidarbha region of Maharashtra, which caters to a large Sickle cell population. The study was conducted in cohort of sickle cell patients. Patients having age more than three years were selected. All patients selected in study are registered patients of our institute and they regularly visit the hospital for consultation and care. Before the establishment of Sickle Cell unit i.e. Jan 2011, all sickle cell patients were receiving blood transfusion without any standard protocol. They were receiving blood on minor pain, mild to moderate anemia, weakness, infection or on any other cause of admission. After its establishment a uniform protocol was followed so as to cut down unnecessary blood transfusion. Blood was given only to treat sudden and severe complication like sequestration crisis, acute chest syndrome and aplastic crisis. Children with repeated pain crisis, stroke and those requiring frequent transfusion were started on hydroxyurea. None of the patients were on chronic transfusion therapy.

For the study purpose patients were divided in two groups, first one before the implementation of protocol (before Jan 2011) and another group after the implementation of protocol (from Feb 2011 to Mar 2015). Average per person-year blood transfusion was calculated in both the groups. Children less than three years of age were excluded from the study. The present study highlights the importance of selective blood transfusion in the patients of sickle cell anemia.

Result and discussion

Table 1: Sex wise distribution of patients

Gender	No. (%)
Male	47 (59%)
Female	33 (41%)
Total	80 (100)

Above table shows that out of 80 patients, 59% of the patients were male and 41% were females. Male to female ratio was 1.4: 1.

Table 2: Religion and caste wise distribution of patients

Religion and cast	No. (%)
Bouddha	40 (50)
Muslims	14 (17.5)
Hindu	23 (28.8)
Christian	01 (1.2)
Not known	02 (2.5)
Total	80 (100)

Above table depicts that 50% of cases were Buddhist, 28.8% were Hindus and 17.5% Muslims were by religion. Among Hindus Banjara community were found most prevalent, 30.4% of Hindus were belonging to Banjara community.

Table 3 – Genotyping of sickle cell disease in studied patients.

Genotyping	No. (%)
SS	44 (55)
Sbeta thal	33 (41.3)
SD Punjab	03 (03.7)
Total	80 (100)

Table 3 shows that 55% of the patients were having homozygotic form (SS) of sickle cell disease followed by Sbeta thal and SD Punjab.

Table 4: BT/Person/year before and after protocol implementation

BT done	Total person-years	BT units	BT/Person/year
Before protocol	591	914	1.55
After protocol	240	88	0.37
Ratio = 1.55/0.37= 4.18 : 1			

Table 4 shows that before protocol there were 591 person-years (total no. of age in years before the protocol) with total 914 unit blood transfusion. Average per person-year blood transfusion was 1.55 blood transfusions/Person/Year. Similar way after implementation of protocol total person-year was 240 person-years with total 88 blood transfusions. Average per person-year blood transfusion was 0.37 blood transfusions per year/per person. It is clearly observed from data analysis that before protocol implementation average BT was 1.55 BT/person/Year compared to 0.37 BT/Person/Year. This means blood transfusion requirement was reduced by approx 75% of previous transfusions. Rate of blood transfusion was 4.18 times more before uniform protocol implementation.

Conclusion-

Large numbers of sickle cell patients are still treated in peripheral centers by with out any sickle cell blood transfusion protocol. The role of transfusion must balance the advantages and disadvantages of this procedure. A bad chronic transfusion programme may be worse than no transfusion. Large nunbers of patients can be prevented from unduly disadvantage of blood transfusion, also it will prevent un necessary misuse of blood. A good programme is hard to conduct with limited resources and expertise available in government setup. There is need to create awareness among all clinicians caring for sickle cell patients about indications of transfusion and hazards of multiple transfusion in these patients.

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