

## AN AYURVEDIC APPROACH FOR THE MANAGEMENT OF IDIOPATHIC ERYTHROCYTOSIS - A CASE REPORT

Radhika. S. Pukale<sup>1</sup>, Harish Bhakuni<sup>2</sup>, Virendra Singh Hada<sup>3</sup>

<sup>1</sup>P.G. Scholar, Final Year, <sup>2</sup>Assistant Professor, <sup>3</sup>P.G. Scholar;  
P. G. Dept. of Kayachikitsa, National Institute of Ayurveda, Jaipur, Rajasthan, India

Email: [radhika.s.pukale@gmail.com](mailto:radhika.s.pukale@gmail.com)

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### ABSTRACT

An erythrocytosis occurs when there is an increased red-cell mass. The causes of erythrocytosis are divided into primary, when there is an intrinsic defect in the erythroid cell, and secondary, when the cause is extrinsic to the erythroid cell. An idiopathic erythrocytosis occurs when the increased red-cell mass has no identifiable cause. Primary and secondary defects can be further classified as either congenital or acquired causes. According to Ayurveda classics IE can be correlated as *Rakta Pradoshaja Vikara* due to *Kapha Dushti* leading to increased *Picchilatwa* (hyper viscosity) in *Rakta*. The present case study deals with chronic case of IE to stop the persistent increase of red blood cells mass & viscosity of blood. In this case, an effort was made to treat 38 years old male patient presenting with headache, pain in heel, weakness, increased RBC mass, increased Hemoglobin % with hyper viscosity in blood, was advised a regular medically prescription of phlebotomy every 8<sup>th</sup> week since 3yrs. This patients was treated with *Shuddh Tamra Bhasma*, *Panchakola Choorna*, *Dashamoola Madhudak*, *Tab. Liv-52DS* orally in BD Dose with *Ushna Jala* as *Anupana*. for a duration of 3months. During the treatment it was observed that patient showed much improvement in his symptoms, reports yielded Hb% & HCT (measure of hyper viscosity) maintained under normal limits.

**Keywords:** *Rakta Pradoshaja Vikara*, *Shuddha Tamra Bhasma*, *Panchkola Choorna*, *Dasamoola Madhu Udak*.

## INTRODUCTION

Erythrocytosis is an increase in the number of red blood cells. In a recent study, the prevalence of primary erythrocytosis (known as polycythemia vera) was 44-57 per 100 000<sup>1</sup>. Erythrocytosis<sup>2</sup> is suspected when hemoglobin is above 18.5g/L or the packed cell volume is greater than 0.52 in a man or 16.5 g/L and 0.48, respectively in a woman. Erythropoiesis is a tightly regulated process by which the bone marrow produces and maintains a normal red blood cell mass (RCM) required to provide adequate tissue oxygenation. The term 'idiopathic erythrocytosis' (IE) applies to a situation when a patient has a measured red cell mass (RCM) above the normal range, manifested as a persistent increase of venous hematocrit levels for 2 months. i.e. an absolute erythrocytosis, but-following further investigation, no cause of primary or secondary erythrocytosis/polycythemia can be established<sup>3</sup>. The condition has been variably called 'benign erythrocytosis' or 'pure erythrocytosis'. The term 'idiopathic erythrocytosis' is preferred because it emphasizes that the underlying cause, driving erythropoiesis is unknown. Most patients with an absolute erythrocytosis have an identifiable cause and can be classified into primary and secondary erythrocytosis<sup>4</sup> types based on the responsiveness of the erythroid progenitors to erythropoietin (Epo). Primary polycythemia is characterized by an autonomous and/or augmented response of the hematopoietic progenitors to Epo due to inherited germ line or acquired somatic mutations; this group includes PV. By contrast, secondary polycythemia has normal responsiveness of the erythroid progenitors to Epo and are due to increased levels of circulating factors driving erythropoiesis. Idiopathic Erythrocytosis constitutes the remaining group of patients in whom no cause for polycythemia has yet been identified.

**Risk factors**<sup>5</sup> Patients at increased risk of occlusion, such as those with evidence of ischemia, previous history of thrombosis, peripheral vascular disease, diabetes mellitus and hypertension.

### Initial Investigations<sup>6</sup>

CBC, Iron studies/ferritin Serum erythropoietin concentration JAK2 mutation, Oxygen saturations, Renal

and liver profile, Abdominal ultrasound, Chest radiography, Lung functions, Bone marrow aspirate. Diagnostic criteria by WHO<sup>7</sup> requires meeting both major criteria and 1 minor criterion or the first major criterion and 2 minor criteria.

**Major Criteria** include Hemoglobin >18.5 g/dl in men, 16.5 g/dl in women, or other evidence of increased red cell volume with Presence of JAK2V617.

**Minor Criteria** includes Bone marrow biopsy showing hyper cellularity for age with trilineage growth with prominent erythroid, granulocytic, and megakaryocytic proliferation, Serum EPO level less than the reference range for normal, Endogenous erythroid colonies formation in vitro

**Treatment** Venesection/ Phlebotomy<sup>8</sup> (usually 450 mL of blood; to maintain a target packed cell volume <0.45), together with Low Dose Aspirin (100 mg daily), is initial therapy that has been recently found to reduce the incidence of major vascular complications According to Ayurvedic literature, while explaining the importance of *Rakta Dhatu*, Acharya Charaka quotes *rakta* as *prana* (vital function) and further enlists *Daurbalya* (weakness), *Agnisaad* (generalized body weakness), *Aruchi* (tastelessness), *Shirashoola* (headache), *Buddhisammoha* (confusion) as classical features of *Shonita pradoshaja vikara*<sup>9</sup>. The *Shuddha Rakta*<sup>10</sup> is responsible for *Bala* (strength), *Varna* (complexion), *Sukha* (comfort), *Ayushya* i.e. enhances longevity (*cha su 24/4*)<sup>11</sup> Acharya Sushruta has mentioned *Rakta* as *Panchabhoutika*, based on which it possess following *Karma*(functions) like *Visrata* (foul), *Dravata* (fluidity), *Raga* (redness), *Spandan* (Pulsation), *Laghuta* (lightness)- (*Su.su 14/9*). Amongst which specifically, the *Drava Guna* which is attributed to *Jala mahabhuta* when vitiates in *Rakta* along with *kapha dosha* & *ama* leads to increased viscosity of blood.

*Shleshma dushta rakta* is *Snigdha* (unctuous), *Sheetala* (cold), *Bahala* (thick/increased volume), *Picchila*(increased viscosity), *Chirasravi* (increased resistance to blood flow) Ref -*Su.su 14/21*; *Tantumadghanam* (increased fibrinogen activity) Ref *Cha.su.24/21* Similarly, the Acharyas have also mentioned, the *Vridhdha*

*shonita dhatu* (increased blood) is vitiated by *kapha* and leads to *Bahalam* (increase in volume), *Picchila* (increased viscous nature), *Ghantwa* (increased density) in *Rakta*. देहस्य रुधिरं मूलं रुधिरैव धार्यते | तस्माद्यत्नेन संरक्ष्यं रक्तं जीव इति स्थितिः ||४४||Su.su 14/44 Hence while planning *Chikitsa* for *Rakta dushti* by *Kapha Dosha* its very essential to protect *Agni* due to *Rakta anavasthitata* and treat the *kapha dosha* with *Na ati Ushna Sheeta*, *Laghu Deepaniya*, *Pachana*, *Lekhana* & *Chedana Guna Dravya* with sufficient leveraging of *vata- kapha* axis equilibrium to avoid *Vata Prakopa* during *Chikitsa*. This case report aims at providing a cost effective and new approach towards successful management of Idiopathic erythrocytosis (IE) with effective and safe Ayurvedic formulation.

### Therapeutic intervention

**Table 1:** Showing therapeutic intervention in a case of IE

<i>Oushadha Kalpana</i>	<i>Matra</i>	<i>Oushadha</i> <i>Kala</i> <i>Vyana</i> <i>Kala</i>	<i>Guna karma</i>
<i>Shuddh Tamra Bhasma</i>	30mgBD		<i>Tikta Kashaya, Ushna, Lekhana</i>
<i>Panchakola Choorna</i>	2gm BD		<i>Ushna, Deepana, Pachana</i>
<i>Dashamoola Madhudak</i>	75ml+10gm		<i>Chedana, Lekhana</i>
<i>Tab.Liv-52DS</i>	2-tab BD		<i>Kaphapittapaha, Yakrit Uttejak</i>

**History of Present illness:** Patient states that he was quite well 5 years back, Since then he has been suffering from *Shirashoola* (headache), *Padashoola* (pain in B/L feet), *Daurbalya* (generalized body weakness) for past 5yrs which is relapsing and remitting in nature. The concerned physician treating his symptoms diagnosed the condition as i.e. based on his complete blood profile and initiated the primary treatment with phlebotomy (450 ml units of bloodletting) along with anti-coagulant therapy with Ecospirin 75mg OD. On consecutive follow ups, the CBC (complete blood count) favoured an increase in the RBC mass along with regular prescribed treatment at the end of every two months, the patient could not get relief completely. As patient was not satisfied, he discontinued the medications. Further, the patient felt increase in the intensity of above-mentioned symptoms for about 2 months, after which he preferred visiting Out-patient department of NIA.

### Case Report

38 years old male patient has attended the OPD in Department of *Kayachikitsa*, National Institute of Ayurveda, Jaipur. The patient had complaints related to Idiopathic Erythrocytosis which included headache, fatigue, pain in heels, weakness with his investigations as mentioned as follows. As in IE, due to continuous increase in RBC counts, hemoglobin (Hb), Hematocrit (HCT) and considering the *Kaphaja Rakta dushti*; *Laghu Deepaniya* (improves metabolism), *Chedana* (dissolving/reducing the fibrinogenesis), *Lekhaniya guna* (scarping action) *yukta oushadi* were selected. These medicines mentioned in Table No 1 act on *Yakrit* & *Pleeha* which is *moola* of *Rakta dhatu* were prescribed.

At the time of presentation, the patient was more concerned with headache, pain in feet, also with consistently increasing blood cell counts, PCV, HCT and its associated risks. Headache was of throbbing type perceived all over head triggered on lying down as well as on changing postures. Headache was associated with feeling of nausea & fatigue. Pain in feet /heels increased on walking & dropping down legs towards gravity. Patient didn't complain about any visual disturbances/vertigo/SOB/Nose bleeds. According to his family he did not snore or have any apneic periods during sleep. There was no history of smoking or any cardio-pulmonary disease and transfusion of blood products. The patient did not have a familial history of erythrocytosis.

**Past history of Patient** The patient had been suffering with similar symptoms for past 5 years, His family history revealed that there was no such pertinent manifestation of disorders. The following findings were observed on evaluation.

**Table 2:** Showing previous Laboratory investigations

2/4/13	<b>USG (whole abdomen) –NAD</b> <b>VIT B12- WNL</b> STOOL/E -NORMAL LAP score -108 PBF-normal	HB-17.0gm% RBC -5.64 million/cumm MCV-86u <sup>3</sup> MCH-30pg MCHC-35% Platelets- adequate PCV-50.6%
3/4/14	<b>2DEcho –LVEF=60%</b> All chambers normal IVS&IAS intact	
26/4/11	<b>EPO-15.5 (range 3.7—29.5Iu/ml)</b> <b>JAK2 Mutation- not detected</b>	
6/11/17	<b>Bone marrow biopsy – Mildly Hyper-cellular marrow with adequate tri-lineage hematopoiesis</b>	

**Roga- Rogi pariksha**

**Probable Causative Factor could be Kshar** (high alkaline food intake), *Amla* (Sour), *Dadhi* (curd), *klin-nabhakshana* (improper food intake habits like frozen food/stale food), *Atapa* (excessive exposure to sunlight & to heat sources), *Adhyashana* (Eating excessive quantity of food frequently), *Shrama* (excessive labor/hard physical work) **and presented premonitory symptoms like** *Ragata* (redness of eyes), *Agnisaada* (Loss of appetite), *Vyahat purishavega* (disturbed bowel habits). The Symptomatology presented as *Santapa* (excessive feeling of hot flushes), *Shirahshoola* (headache), *Padashoola* (pain in heels), *Atidaurbalya* (excessive weakness), *Tamaso darshana* (blackouts), *Buddhi sammoha* (generalized anxiety). The symptoms aggravated on exposure to *Atapa* (sunlight/ heat radiating sources) which is *anupashaya* and were observed by the patient to be decreased on *rakta avasechana* (phlebotomy)- *upashaya*.

**Samprapti ghataka**

The predominance of *doshas* found as per symptomatology were observed to be due to *Ati-guru, Picchila guna* of *Vikruta Kapha dosha* along with *prana-vyana vayu vikrati* (abnormal functioning of *prana vayu*) leading to *Sanga* in *Rakta vaha srotas*. These *vikruta dosha sanga*, on further chronicity leads to *khavaigunya* in *rakta dhatu* due to improper functioning of *rakta dhatvagni*. The site of *Udhbhavasthana* (initiation of pathogenesis) was retracted to *koshtha marga (yakrit, pliha)* which is *moola* (primary site) for

*raktavaha srotas*, disturbing the normal equilibrium of homeostasis. The cascade of pathogenesis is *chirakari* (prolonged) further progressing to *sarvangashareera* (throughout body with multisystem involvement) leads to manifestation of symptoms like *Santapa* (excessive feeling of hot flushes), *Shirahshoola* (headache), *Padashoola* (pain in heels), *Atidaurbalya* (excessive weakness), *Tamaso darshana* (blackouts), *Buddhi sammoha* (generalized anxiety).

**Astavidha rogi pareeksha**

*Nadi was guru* (heavy with high volume) and *Pittakaphaja* in nature, *Mala was Saama, Shushka*, with difficulty in passing and incomplete evacuation. *Mutra* (urine) was *prakrit* (normal quantity and frequency). *Jihwa* (tongue) was found to be *Shwetabha* (whitish) and *Malavrita* (coated). Patient was having normal body temperature but warm on touch (*Ushna Sparsha*) and his skin texture was *Snigdha* on touch. *Shabda* (voice quality) was *Ruksha* (rough). Examination of the *Drik* (eye) revealed *prakrit drushti* whereas *Akriti* (built) was *madhyam* (normal).

**Personal history of patient:** Appetite - Decreased; Bladder- Clear, Sleep- Normal; Addiction- None; Bowel- irregular, Diet- vegetarian

**Clinical Examination**

During examination patient was conscious, cooperative, fully oriented with intact short term and long-term memory. Patient exhibit his BMI – 19.6kg/m<sup>2</sup> (height - 175cm, weight - 60kg). Patient's pulse was *Kaphavata* dominant with a rate 76beats/min, regular rhythm and

high volume. Supine blood pressure was 130/80mmHg and in upright posture was 120/80mmHg. Respiration was thoraco-abdominal in nature with rate 20/min. Patient was having mild pallor, with no cyanosis, icterus, clubbing and dependent edema. Thyroid and different group of lymph nodes were not palpable. Cardio-Respiratory system examination was fairly normal with vesicular breathing. S1 & S2 were clearly audible with

normal rate and regular rhythm. There were no murmurs or gallops. During vascular assessment all peripheral pulses were palpable with no signs of poor peripheral circulation, no arterial bruits as well as venous hum were present. Neurological assessment showed intact superficial and deep sensation. On motor examination nutrition, tone, power was found to be normal and deep tendon reflexes.

## Timeline

**Table 3:** Shows Timeline of Events Before and After Treatment

Visit	Date	Complaints	Oral medication
1	2/4/19	Loss of appetite /pain in heels with weakness (when RBC /Hb increases)	1) <i>Shuddh tamra bhasma 30mg Panchakola choorna 2 gm / BD</i> 2) <i>Dashamoola madhudak (15gm dashamoola +400ml water remains till 75 ml + 10 gm honey)</i> 3) Tab.Liv-52DS
2	23/4/19	Relief in previous symptoms	1) <i>Shuddh tamra bhasma 30mg Panchakola choorna 2 gm / BD</i> 2) <i>Dashamoola madhudak (15gm dashamoola +400ml water remains till 75 ml + 10 gm honey)</i> 3) Tab.Liv-52DS

## Results

**Table 4:** Showing improvements in Investigations

Sr.No	Date	Complete Blood Count
1	19/2/19	Hb -16.7gm% RBC-6.10million/cumm Hct -54.1 % MCV-85.4 fl MCH-27.4 pg/ml MCHC-32.1 g/dl Basophils-0.8 %
2	22/4/19	RBC-5.76 million /cumm Hb -15gm% MCV-82.8fl MCH-26.0 pg/ml MCHC-31.4 g/dl Hct -47.7% Basophils 0.6%

There was marked and rapid improvement in his symptoms and wellbeing without phlebotomy & anticoagulant therapies in duration of two months. There was marked and rapid improvement in following investigations as shown in Table no 3

## DISCUSSION

In this study, due to hyper viscosity i.e. *Picchilata* (viscous) and *Ghanatwa* (increased density) of *Rakta* is due to *Kapha dusti* in *Raktadhatu*. The treatment protocol followed was *na ati ushna sheeta, Laghu Deepaniya*



and hence the drugs needed to treat this must have, *Pachana* (enhancing proper metabolism), *Lekhana* (scraping) & *Chedana* (dissolving clots) *guna*. So, the choice of drug was *Shuddha tamra bhasma 30mg* with combination of *Panchkola choorna in Vyana kala with Ushna jala anupana*. According to *Ayurveda Prakash & Rasamrit*<sup>12</sup> *Shuddha Tamra* (purified copper) is considered as *Tikta-kashaya rasa* (astringent), *Ushna veerya*, *Madhura vipaka*, *Laghu*, *Lekhana & Saram*. These properties are *Pittakapha hara*, also acts on *Rakta dhatwagni* normalizing the *Prakrit karma* of *Rakta dhatu*. According to *Rasachudamshu*, *Shuddha tamra* is considered as “*Param Lekhaniyam*” (scraping effect), due to which the risk of thrombotic events was reduced down and *ushna veerya* helps in normalizing the hyper-viscous nature of blood.

*Panchakola churna* consists of *Pippali* (*Piper longum* Linn.) *Pippalimula* (Root of the *Piper longum* Linn.), *Chavya* (*Piper chaba* Hunter), *Chitraka* (*Plumbago zeylanica* Linn.) and *Sunthi* (*Zingiber officinale* Roscoe) are the ingredients of *Panchakola Churna*. All the ingredients have *Laghu*, *Ruksha* and *Sukshma Gunas*, *Ushna Virya*, *Katu Rasa*, *KatuVipaka* and *Pachana*, *Kaphahara* properties. *Rasa* and *Vipaka* of the *Panchakola* are *Katu*, helpful in *Agnidipana* and *Pachana*. Thus, it becomes clear that *Deepana & kaphahara* properties correct *Agnivaishamy* and *Doshavikriti*. After the proper channelization of *Rasadi Dhatu Samvahana*, *Rakta vikriti* with *kapha* was checked maintaining *Swasthya* which entitles “*Samagni, Samadhatu*” (maintaining health) axis. Liv.52<sup>13</sup> DS Tablet is manufactured by Himalaya Drug Company and it contains a number of active ingredients including *Himsra* (*Capparis spinosa*), *Kasani* (*Cichorium intybus*), *Mandura bhasma*, *Kakamachi* (*solanum nigrum*), *Arjuna* (*Terminalia arjuna*), *Kasamarda* (*Cassia occidentalis*), *Biranjaisipha* (*Achillea millefolium*), *Jhavuka* (*Tamarix gallica*) acts on *yakrit* and *pliha* which is *rakta vah srotas moola* and is responsible for *Pachana* (metabolism & excretion) of *Ama*(toxins) leading to *Kapha shaman*, enhancing the *Prakrit drava* (fluidity), *sara* (free flowing nature) *guna of rak-tashrit pitta*. *Dashamoola Madhu udaka - Dasha-*

*moola*<sup>14</sup> is useful herbal combination of *laghu panchamoola i.e. Gokshura* (*Tribulus terrestris*), *Shalaparni* (*Desmodium gangeticum*), *Prishniparni* (*Urariapicta*), *Kantakari* (*Solanum xanthocarpum*) and *Bruhati* (*Solanum indicum*) and *Bruhat panchamoola i.e. Bilva* (*Aegle marmelos*), *Agnimantha* (*Premna integrifolia*), *Shyonak* (*Oroxylum indicum*), *Gambhari* (*Gmelina arborea*) and *Patala* (*Stereospermum suaveolens*). *Laghu panchamoola* indicated for *VataPitta* disorders and *Bruhat panchamoola* is indicated in *Vata-Kapha* disorders. *Dashamoola* is *Vatananuloma*, *Tridosha-nashak Madhu* (honey) is *Ruksha*, *Kashaya*, *Chedhana*, *Sukshma marga anusari*, *Ropana*, *Raktapittakaphapaha*. *Yogavahi guna* is enriched with enzymatic properties helps in enhancing the effects of *Dashamoola* increasing *Lekhana karma*. Due to the *Grahi* effect of *Dashamoola* the *Prakshepa* of *Madhu* in it will help to counteract the effect and acts as *Amapachana* (enhances metabolism), *Bhedana* (dissolving the hyperdensity & hyper viscosity), *Anulomana* (normalizes the functions of *Vayu*).

## CONCLUSION

Marked improvement was found in all the signs and symptoms of the disease. After completion of treatment there is maintenance of proper blood counts without any complication or adverse effects and was beneficial in terms of cost effectiveness. It is an effort to understand the concept of *Shonitapradoshaja vikara* w.s.r. to *Kaphadusti* in *Rakta* with a view of applied aspects of Idiopathic erythrocytosis.

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