

Table (2) literature review summary

Reference	Age	Gender	Nationality	Clinical presentation	Comorbidities	Family history	Hemoglobin	Methemoglobin	Type	Management	Follow up
6	2 days	Male	NA	Cyanosis	None	Mother had cyanosis, methemoglobin 14%	28 gr/dl	4%	1	None	Follow up at age 6 months, normal development
7	18 years	Male	Bangladesh	Cyanosis Low exercise tolerance	None	None	18 gr/dl	36%	1	Ascorbic acid	Improved
8	55 years	Male	NA	Cyanosis, flu-like symptoms	CKD, thyroidectomy due to thyroid cancer AF	None	NA	15%	1	Ascorbic acid	Improved
9	61 years	Male	China	Cyanosis erythrocytosis	High altitude	Parents and one brother have cyanosis	22 gr/dl	44.2%	1	Methylene blue	Improved
10	29 years	Male	India	Cyanosis and fatigue	Diagnosed as PV and started imatinib	None	20 gr/dl	38%	1	Ascorbic acid	Improved
11	36	Female	Japanese	Cyanosis, convulsions	None	None	NA	33%	1	Ascorbic acid	Improved
12	24 day	Male	NA	Cyanosis	No consanguinity	Mother cyanotic	20 gr/dl	26%	1	Methylene blue and ascorbic acid	Improved, normal growth and development
13	61 year	Female	Turkish	Mild cyanosis	None	Daughter	NA	16%	1	Ascorbic acid	Improved
13	17 years	Female	Turkish	Mild cyanosis	None	Mother	NA	12.7%	1	Ascorbic acid	Improved
14	40	Male	India	Flu-like symptoms	None	NA	18.5 gr/dl	38%	1	NA	NA
15	18 years	Male	India	Cyanosis, headache, bilateral ptosis	None	Sister	NA	37%	1	Ascorbic acid, aspirin	Improved
16	20 years	Male	NA	Chest pain, exertional dyspnea, cyanosis	None treated as asthma	Brother	NA	40%	1	Methylene blue	Level improved to 8%, cyanosis and other symptoms

											improved
17	21 years	Male	India	Mild cyanosis and exertional dyspnea	None	None	NA	29.5%	1	No treatment	Mild cyanosis
18	16 years	Male	Arabic	Cyanosis, fatigue, shortness of breath on exertion	None	Brother	17 gr/dl	40%	1	Oral vitamin C	Improved
18	12 years	Male	Arabic	Asymptomatic	None	Brother	NA	21%	1	None	NA
19	55 years	Male	India	Cyanosis, weak, syncope	HTN, smoker	None	15 gr/dl	30%	1	Vitamin C, riboflavin	Improved
20	33 years	Male	India	Cyanosis	None	None	18 gr/dl	20-30%	1	NA	NA
21	64 years	Male	Italian	Marked cyanosis	None	Positive	NA	36%	1	NA	NA
22	1 month	Male	India	Central cyanosis	None	Mother	14 gr/dl	29%	1	Methylene blue	NA
22	32 years	Female	India	Recurrent early pregnancy loss	None	Positive	14 gr/dl	7.5%	1	No indication	NA
22	34 years	Male	India	None	None	Positive	15 gr/dl	2.8%	1	No indication	NA
23	NA	Female	NA	NA	NA	Positive	NA	NA	1	NA	NA
23	NA	Male	NA	NA	NA	Positive	NA	NA	1	NA	NA
23	NA	Male	NA	NA	NA	Positive	NA	NA	1	NA	NA
24	2 years	Female	Spanish	Neurological symptoms	NA	Positive	NA	NA	2	NA	NA
25	NA	Male	Japanese	Mental retardation, skeletal anomalies	NA	None					
26	5 years	Female	India	Cyanosis, cerebral palsy, microcephaly, generalized hypertonia	NA	Positive	11 gr/dl	51%	2	Methylene blue Followed by vitamin C	Relapse when stopped vitamin C
26	12 years	Male	India	NA	NA	Positive	9 gr/dl	13%	2	NA	NA
26	8 years	Male	India	NA	NA	Positive	14 gr/dl	42%	2	NA	NA
27	1 year	Female	India	Severe cyanosis, global Developmental delay, generalized	NA	NA	NA	24.7%	2	High dose vitamin C	Cyanosis improved

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**NA: not available, HTN: hypertension, PV: polycythemia vera, CKD: chronic kidney disease, AF: atrial fibrillation**