- Iruone, Aroma Coast, Rigo District
- Bus fare K30/person return
- Born 2nd February 2010
- Age: 10 yrs 6 months
- Dad a private entrepreneur & mum a housewife
- First sibling had Rheumatic Heart disease
- Migrated with parents to Port Moresby in 2010
- Parents are 2nd cousins
- Related to thalassaemic Patients: Patty Dakwana & Tamara Kepere
- She was first transfused at 9/12 old
- Been admitted more than 6 times to Paediatric wards
- Highest education elementary 1-Kalai Primary 2016
- Poor school attendance and never continued.
- Disabled
- On monthly blood transfusions with 2 pack cells
- She is allergic to hydrocortisone
- Parents doesn't want her to know that her cousin Tamara had passed on already..

TIED FOR LIFE

A REVIEW OF THALASSAEMIA PATIENTS RECEIVING BLOOD TRANSFUSIONS IN PMGH BETWEEN FEBRUARY AND AUGUST, 2020."

Violet Regina Nagiria

Masters candidate for Child Health, 2020.

BACKGROUND

- 85% of the world's six billion people live in middle- and low-income countries (UNICEF, 2003)
- 7.9 m — 6% of total births world widely with B.D (MOD 2006). 3.3m children < 5 yrs. of age die from B.D/yr .Est. 3.2m of those who survive may be disabled for life. (MOD Global report on B.D)
- Top 3rd common serious B.D is HbGD: 308, 000 children are born annually.
- HbGD contribute to 3.4% of mortality in children aged < 5 years worldwide or 6.4% in Africa.
- Originally endemic in 60% of 229 countries, potentially affecting 75% of births, now common in 71% of countries among 89% of births to require policy-makers to consider the most appropriate strategy for treatment and prevention.
- Thalassaemia is most common autosomal recessive HbGD on worldwide. 22, 989 are born annually with B thalassaemia major
- > 200 mutations for β-thal. 3% of the world's population carry genes for β-thal & 20% carry α+ thalassaemia.
- 5-10% of SEA poptn carry genes for α-thalassemia including PNG. (1:20)
- Parental consanguinity increases the birth prevalence of autosomal recessive birth defects. The risk for neonatal & childhood death, intellectual disability and serious birth defects is almost doubled for first cousin unions (Bittles et al, 1991; WHO, 1996)

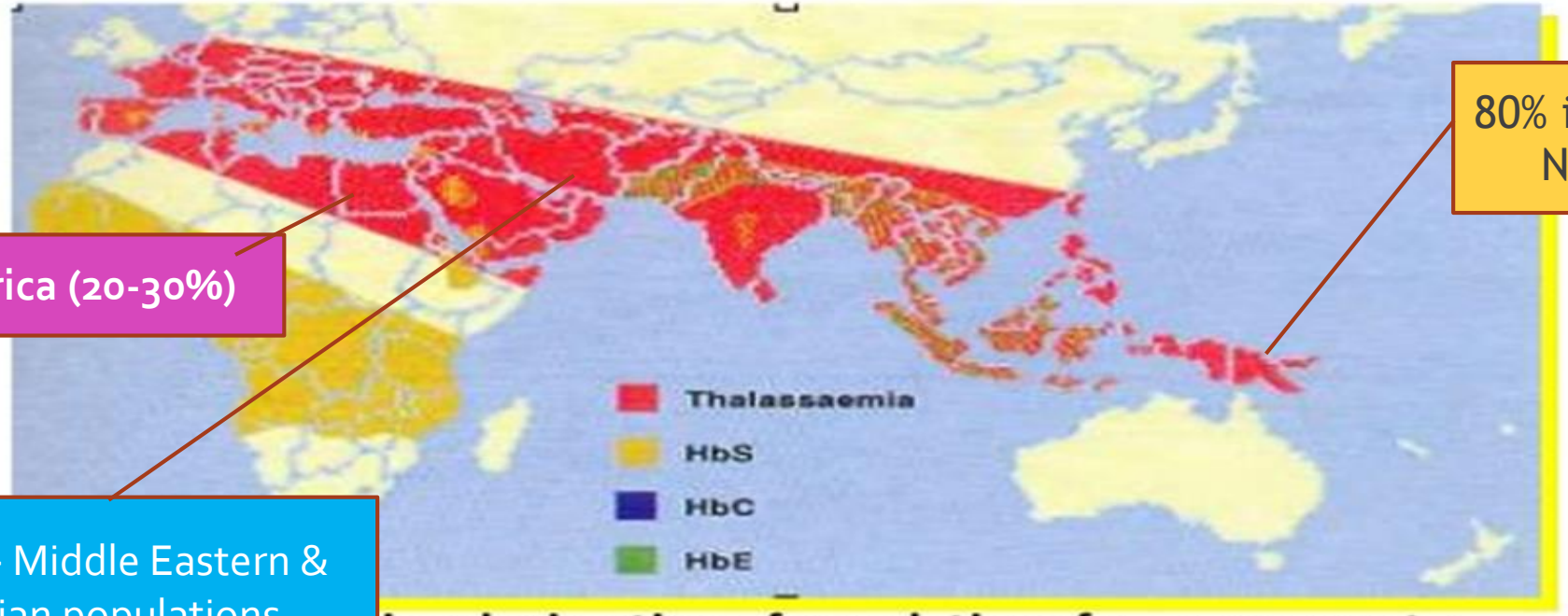
(Modell & Darlingson ;Clegg & Weatherall, 1999; Modell and Kuliev, 1989; Mokenhaupt et al., 2004; WHO, 1996).

INTRODUCTION CONT….

- > 332 000 affected conceptions or births yrly.
- 56 000 have a M.thalassaemia, incl.≈ 30 000 who need regular transfusions to survive & 5500 die prenatally due to α thalassaemia major.
- Most births, 75%, are in countries where HbD are endemic & 13% occur where t is common because of migration, so in principle, 88% of the 128 million women who become pregnant annually should be offered screening.
- > 9 million carriers become pregnant yrly. The risk that their partner is also a carrier ranges from 0.1–40% (global average 14%). In principle, all need information and the offer of partner testing.
- Annually there are at least 948 000 new carrier couples, & > 1.7 million pregnancies to carrier couples. ≈ 75% are actually at risk. In principle, all need expert risk assessment & genetic counselling.
- 1.33 million yrly at-risk pregnancies. In principle, all need the offer of prenatal diagnosis.
- 12% of children born with transfusion-dependent β thalassaemia are actually transfused, & < 40% of those transfused obtain adequate iron-chelation therapy.
- 100 000 patients are currently living with regular transfusions, and at least 3000 die annually in their teens or early 20s from uncontrolled iron overload.

WHO Bulletin:Global epidemiology of Hb disorders & derived service indicators.(Bernadette Modell a, M Darlison)

GLOBAL THALASSEMIA BELT



Sub Saharan Africa (20-30%)

80% in northern PNG & North East India

40% > Middle Eastern & Indian populations

inual migration of populations from one area to another, there is virtually no country of the world now in which thalassaemia does not affect some percentage of the inhabitants

B Thalassaemia Major carrier rate (1-20%)

Alpha Thalassaemia – Milder forms

AIM

- Identify the children diagnosed with thalassaemia & collect their demographic data's
- Discuss the treatment, quality of life and difficulties faced by the children & their families.
- Extract the patients or the guardians view on their experiences and what they think about their child's illness in general and our management & any other views.
- Determine if the thalassemic patients had been receiving optimal care with limited resources available in PMGH.

OBJECTIVES

- Determine how the Thalassemia diagnosis was made.
- Assess:
  - ✓ Age of transfusions and how often
  - ✓ Clinical signs and symptoms
  - ✓ Effectiveness of blood transfusion (Hyper transfusion)
  - ✓ Parents understanding of their child's condition
- Determine the costs per transfusion and the parents or guardians source of income
- Find out the parents or the child's view on life in general and stigmatization
- Assess the effect of thalassaemia on education and general quality of life(QOL).

METHODOLOGY

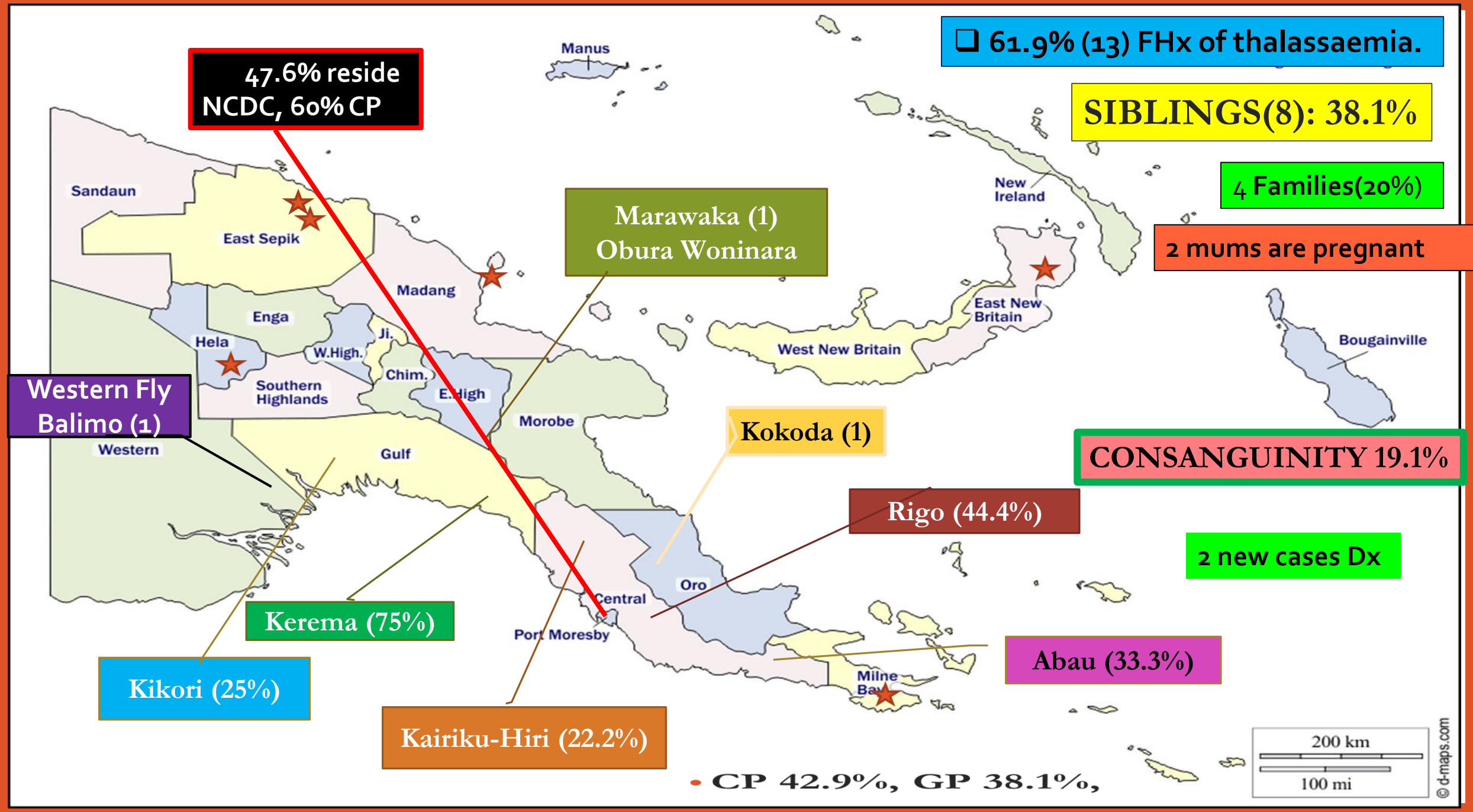
- A descriptive longitudinal qualitative study
- Site: PMGH Children's Emergency Department & Consultation clinic.
- Duration: march 1st – August 31st 2020 (6 months)
- Inclusion criteria:
    - ✓ Any children with Thalassaemia.
    - ✓ Parental and adolescent child consented as well
- Structured questionnaire for parents and children
- Physical assessment and history taking

METHODOLOGY CONT....

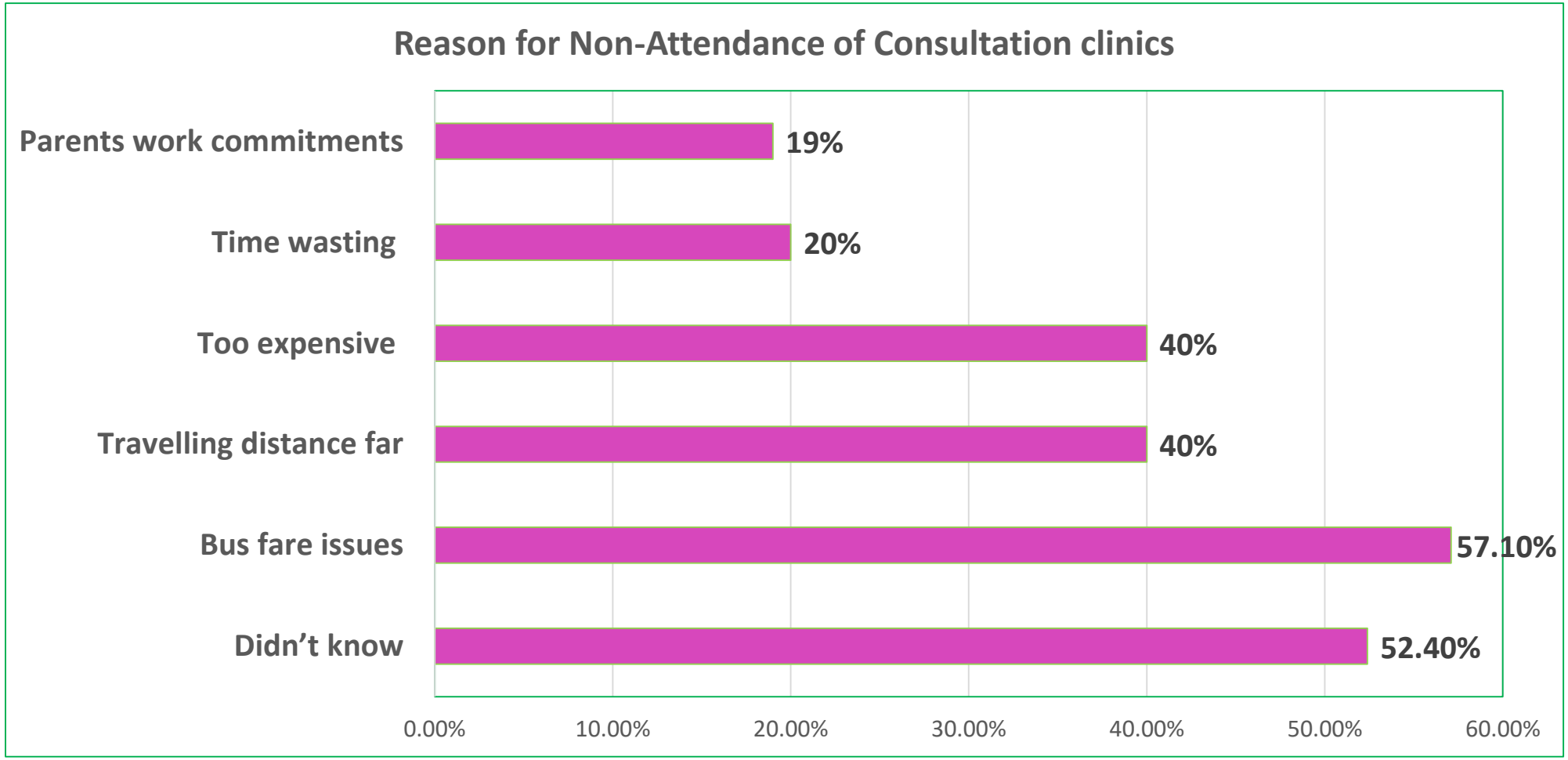
- Follow up weekly for a month then monthly followed by 3/12 then 6/12 etc..
  - ✓ Monitor their regular blood transfusion routine
  - ✓ Explain the genetics to parents
  - ✓ Check on their illness and their perceptions
  - ✓ School attendances and their quality of life
  - ✓ Attend to any complications management with any other social activities
- Get approval for the research from the SMHS Research & Scientific Committee
- Data entry and analysis: Microsoft Excel

RESULTS

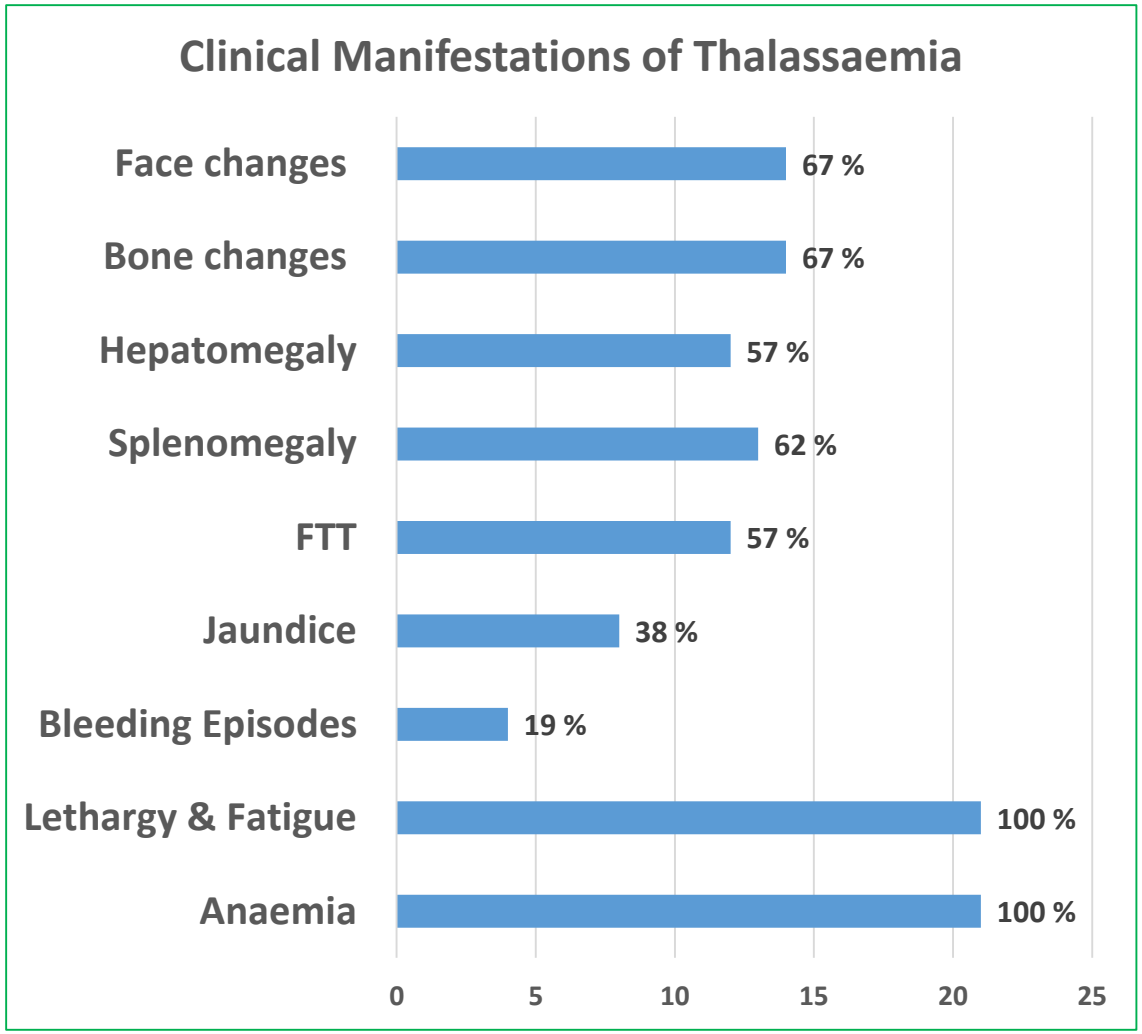
- **Total Patients**: 21
- 68.9% were Female (Gender ratio (M:F) 1- 1.63≈ **1:2**
- Median age: **7.7 years** (IQR: 4.7 – 14.4 (9.7)) – (1yr 1/12 – 21.7 yrs)
- 57.1% were delivered in a Health facility
- 83.3% were born at PMGH
- 1 died (4.8%) at the age of **18 years 7/12**
- Province: **CP 42.9%, GP 38.1%,** Others 14.3% & UK 1 (4.8%)

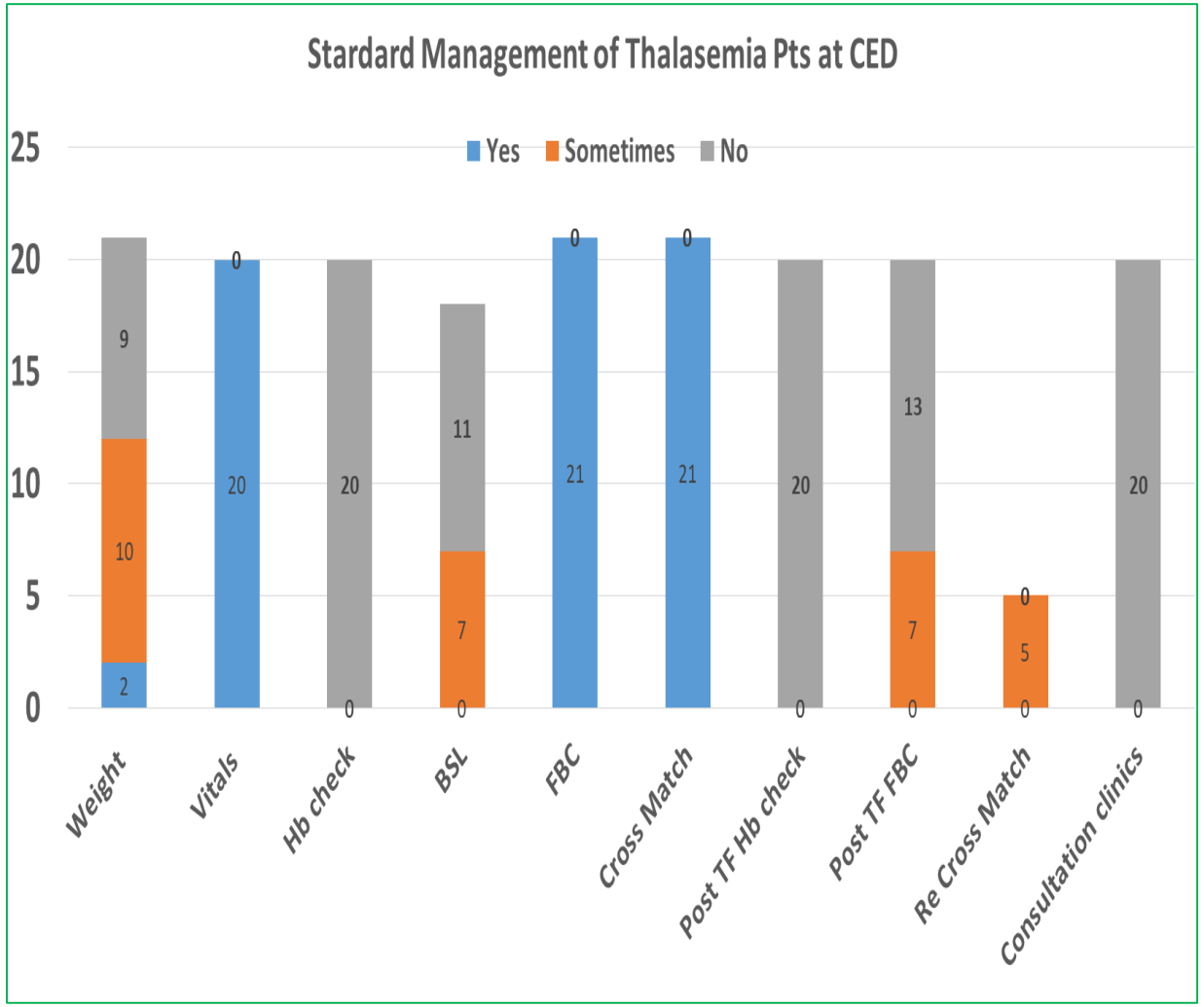

❑ 61.9% (13) FHx of thalassaemia.
47.6% reside NCDC, 60% CP
SIBLINGS(8): 38.1%
4 Families(20%)
2 mums are pregnant
Marawaka (1)
Obura Woninara
Western Fly
Balimo (1)
Kokoda (1)
CONSANGUINITY 19.1%
Rigo (44.4%)
2 new cases Dx
Kerema (75%)
Kikori (25%)
Kairiku-Hiri (22.2%)
Abau (33.3%)
• CP 42.9%, GP 38.1%,
Manus
New Ireland
Sandaun
East Sepik
Madang
Enga
Hela
Ji.
W.High.
Chim.
E.High
Southern Highlands
Western
Gulf
Morobe
East New Britain
West New Britain
Bougainville
Oro
Central
Port Moresby
Milne Bay
200 km
100 mi
© d-maps.com

RESULTS CONT....



RESULTS CONT....





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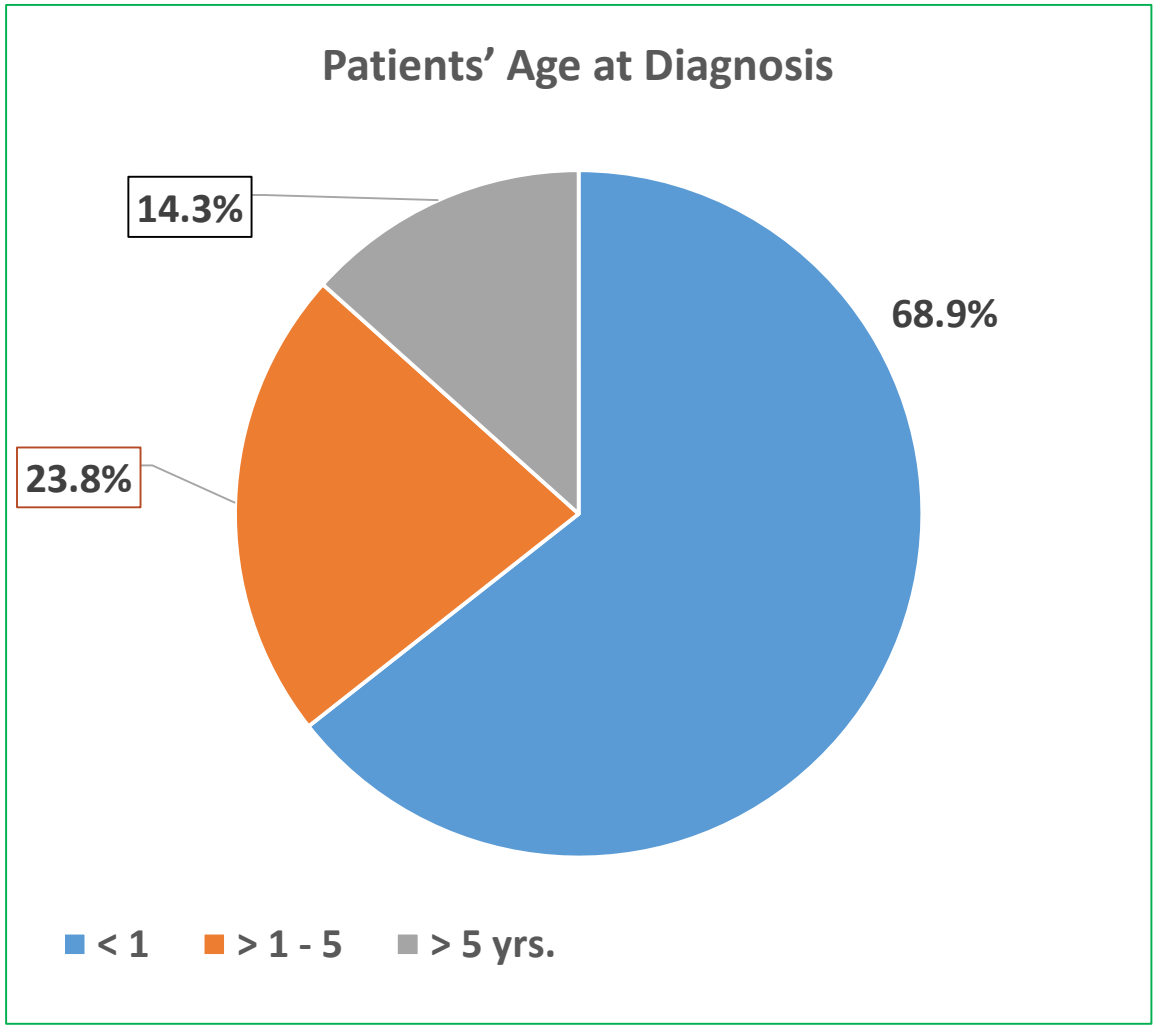
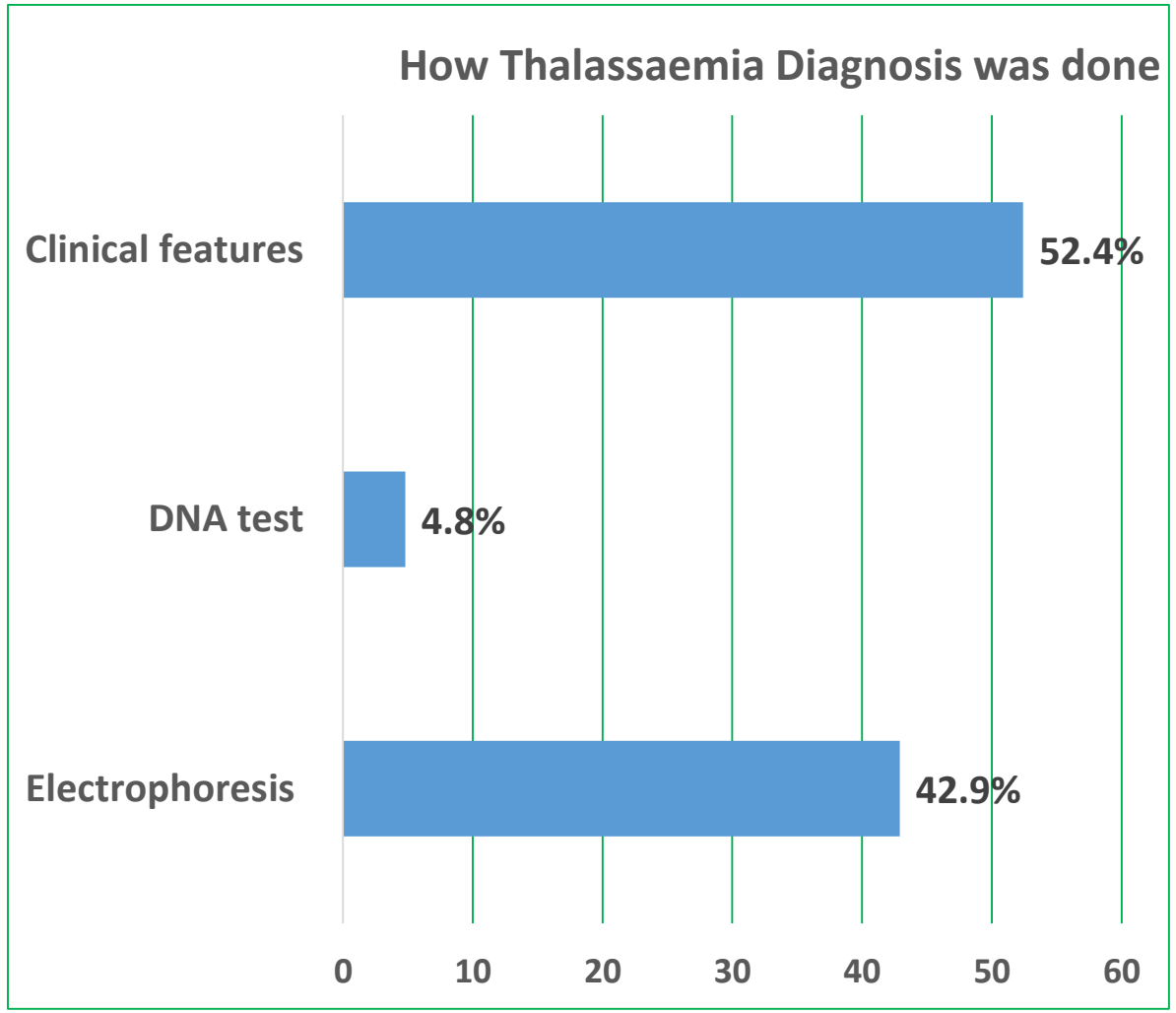
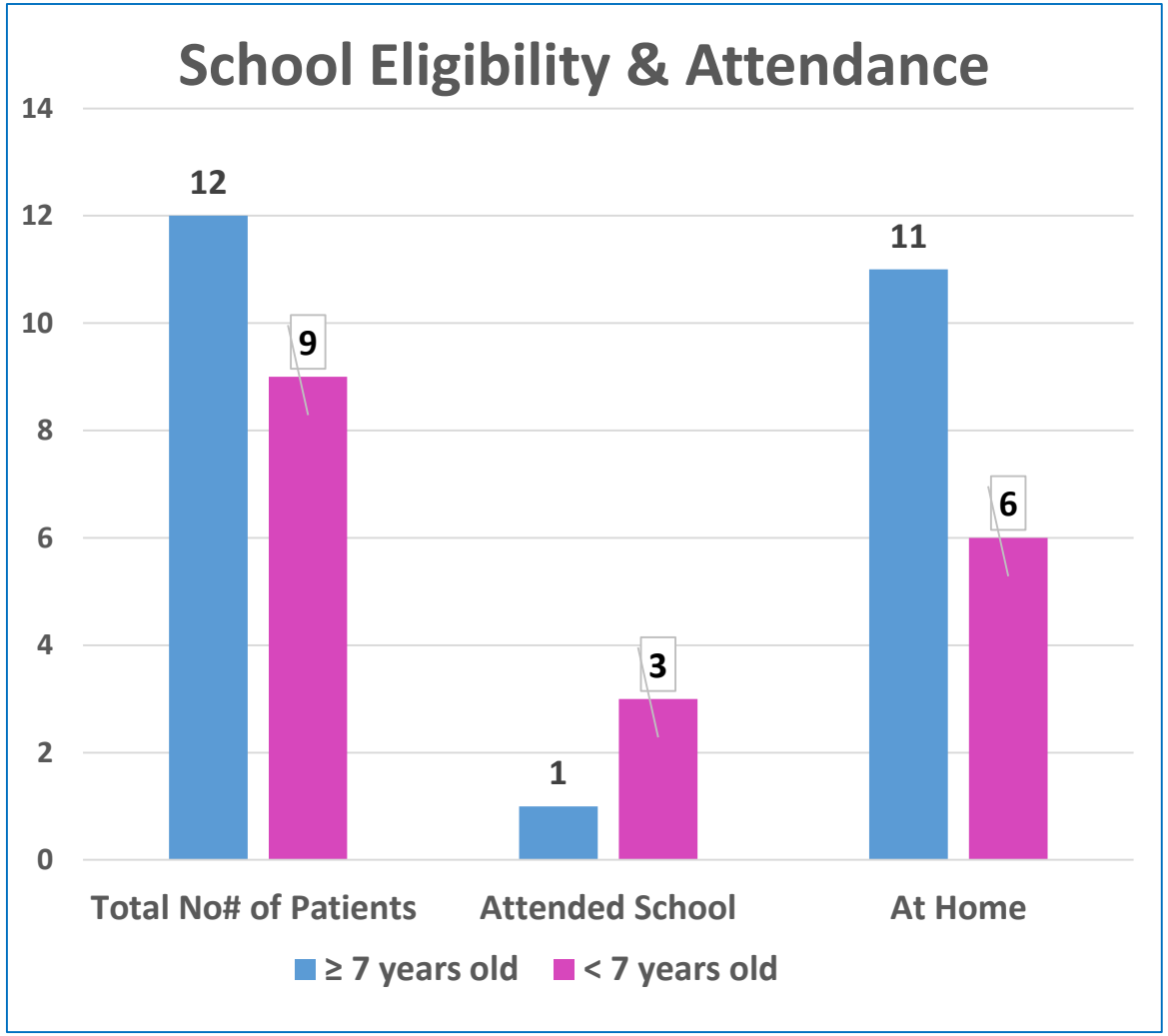
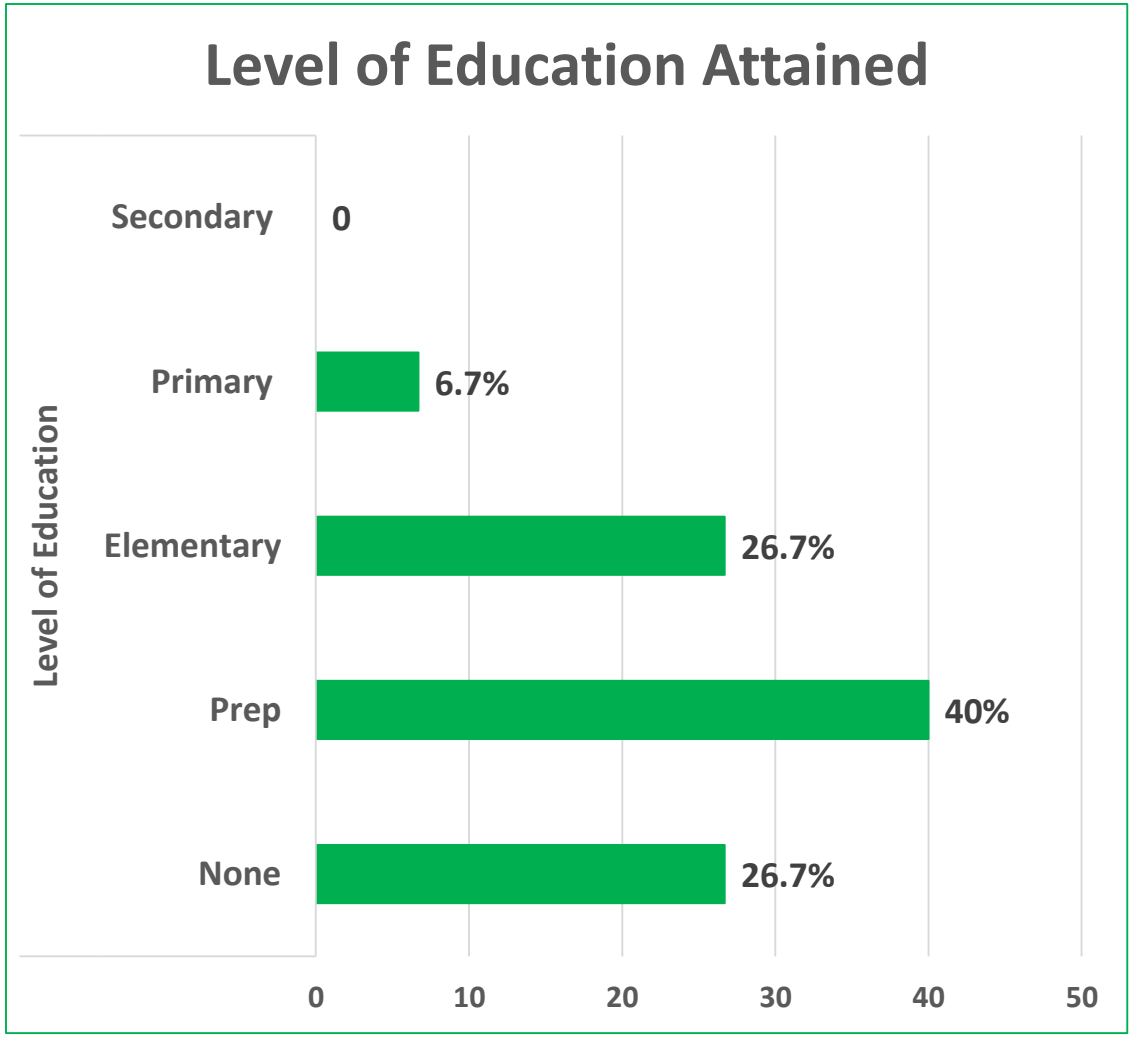


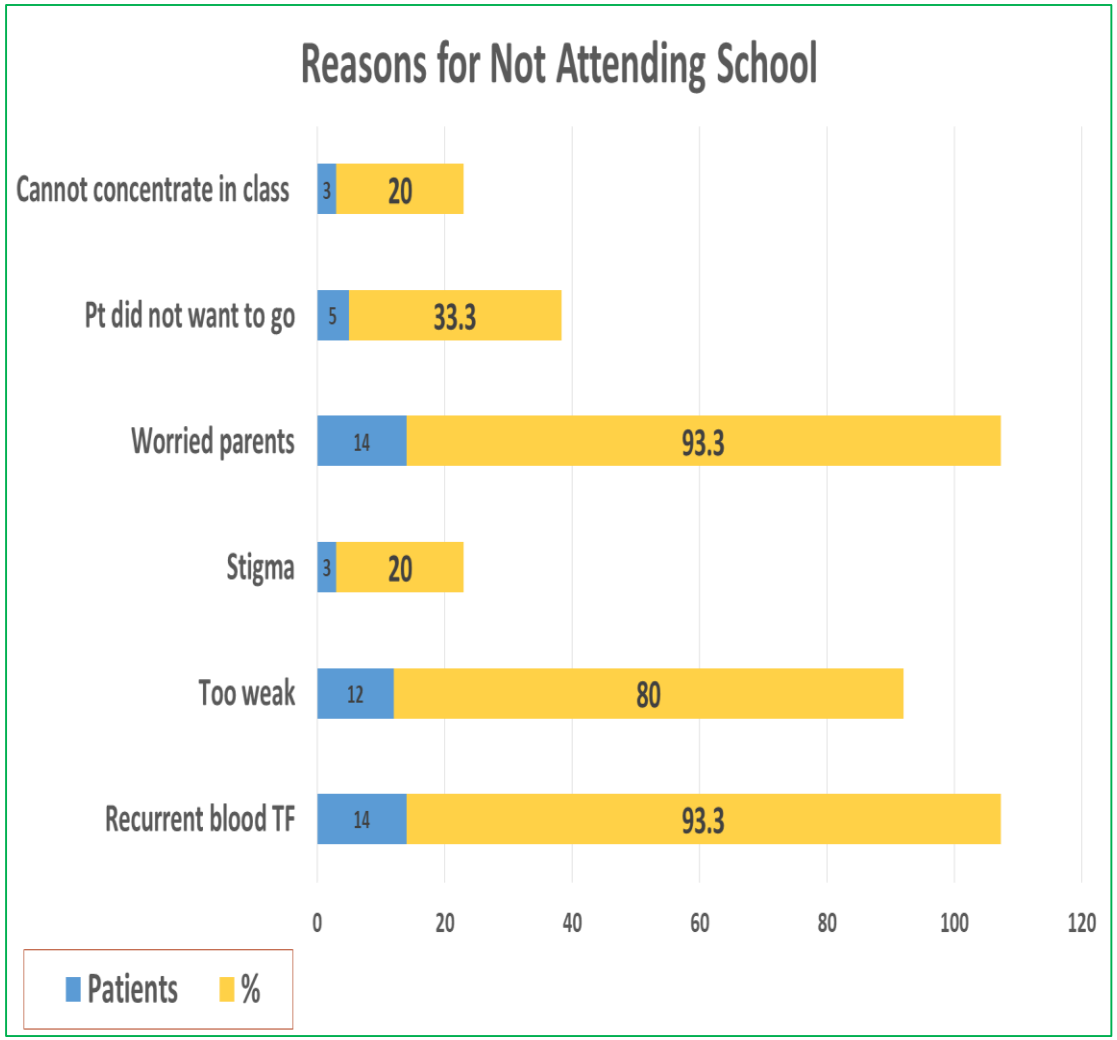


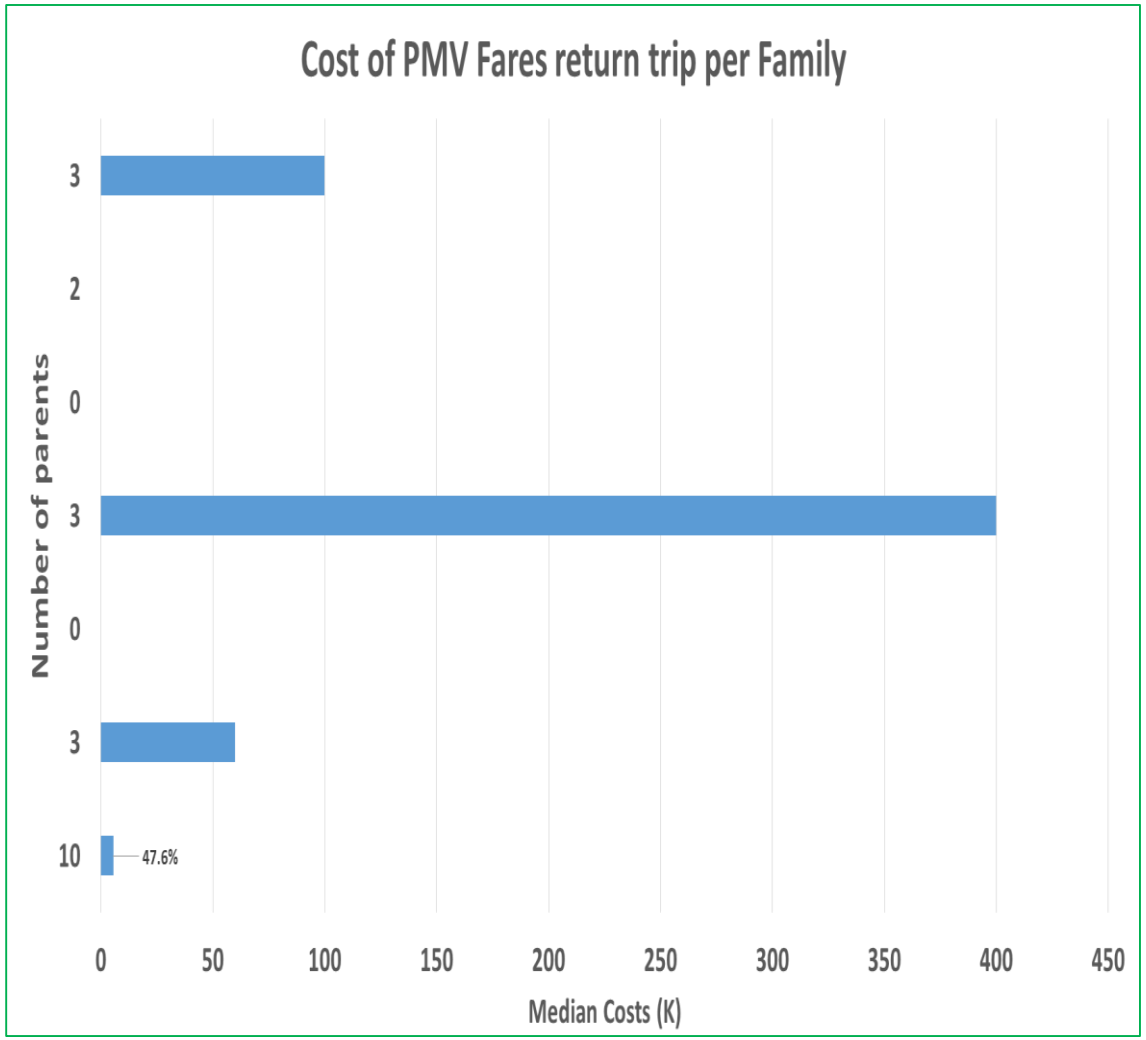
RESULTS CONT....



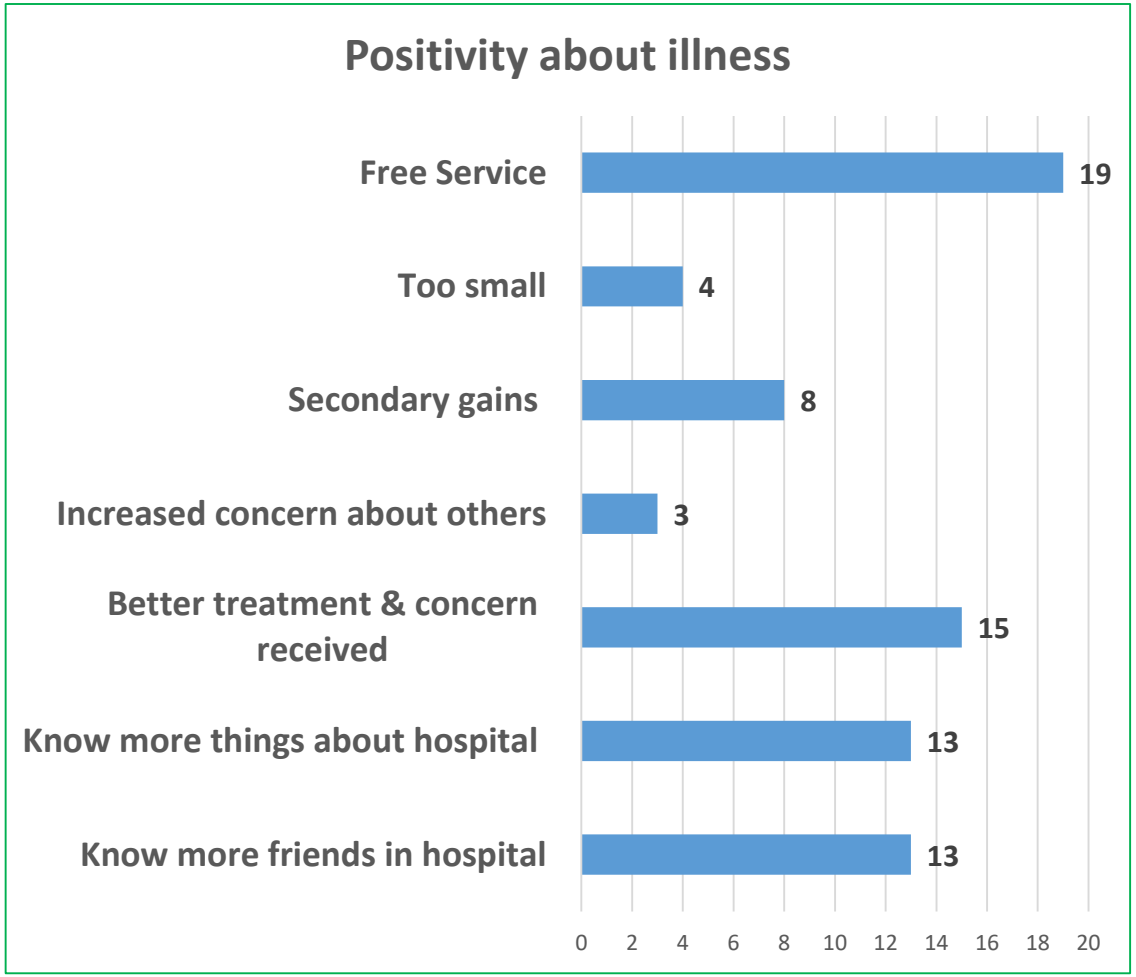


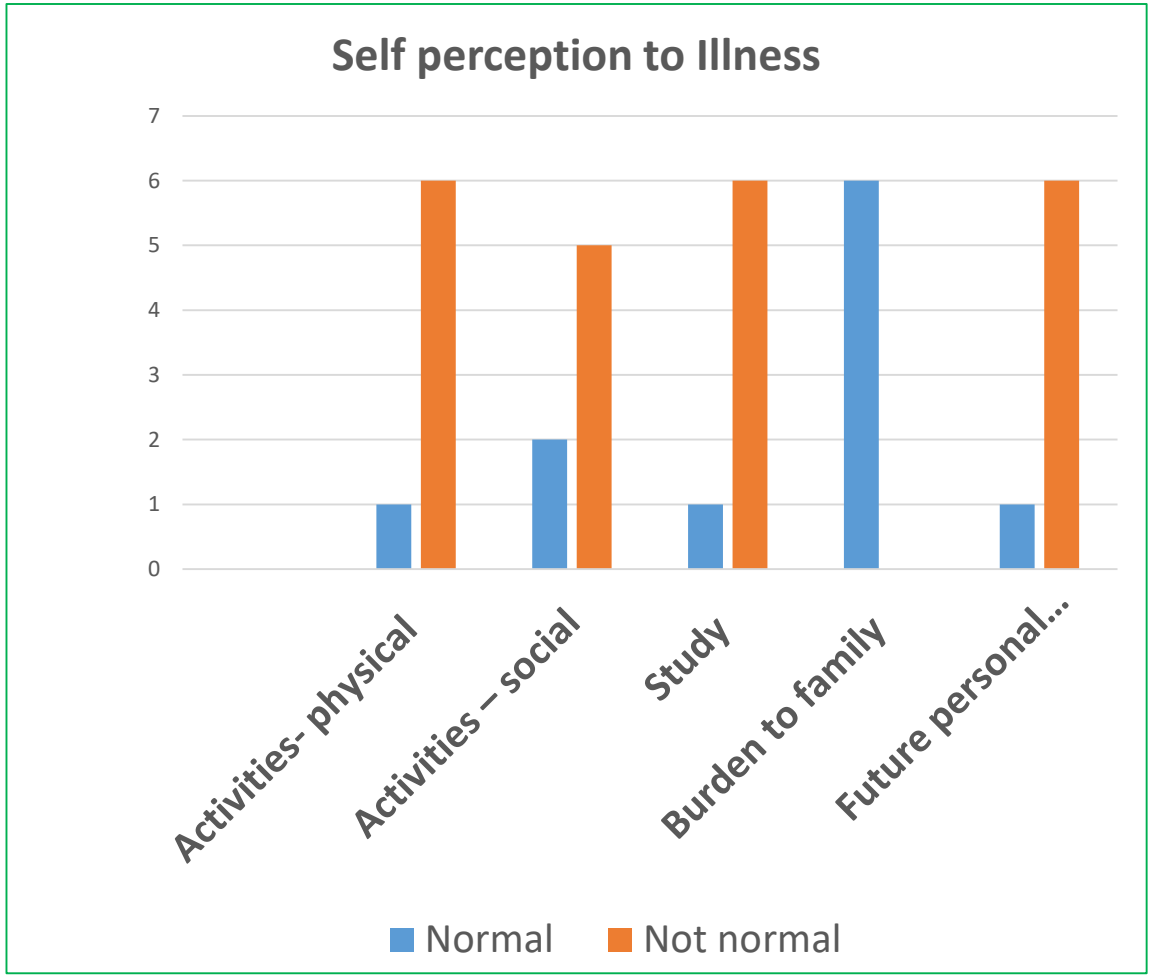
RESULTS CONT....





RESULTS CONT....





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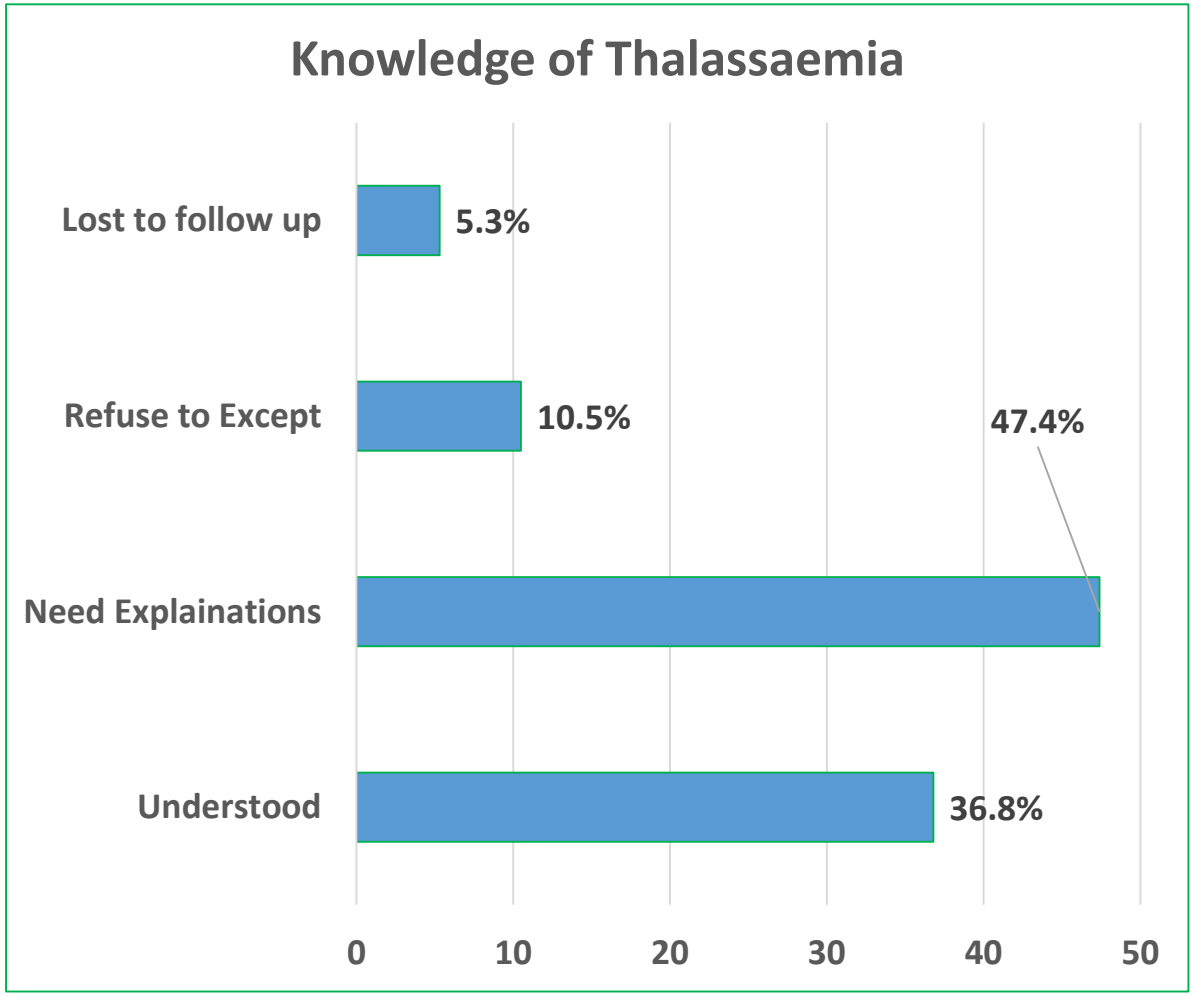
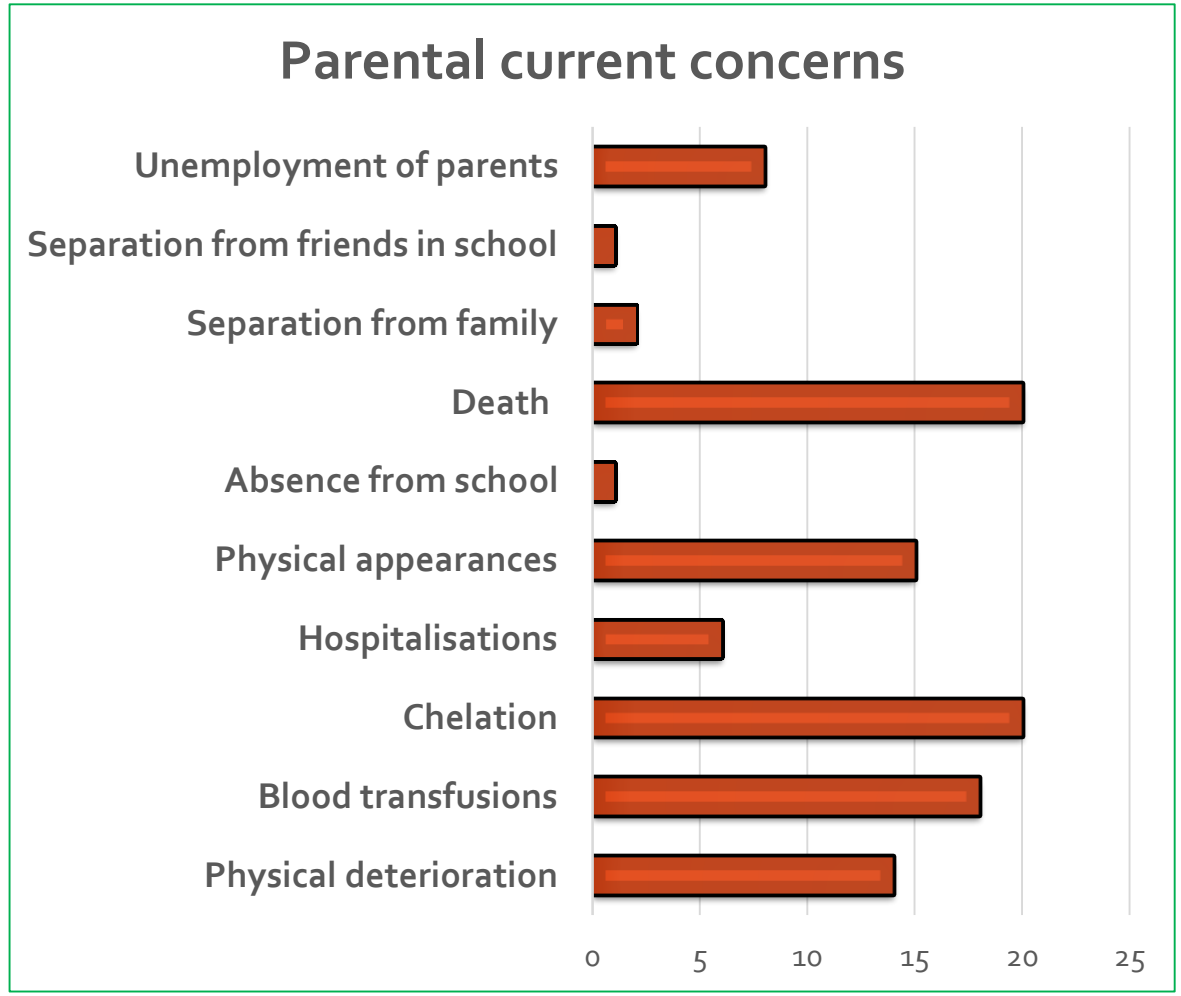
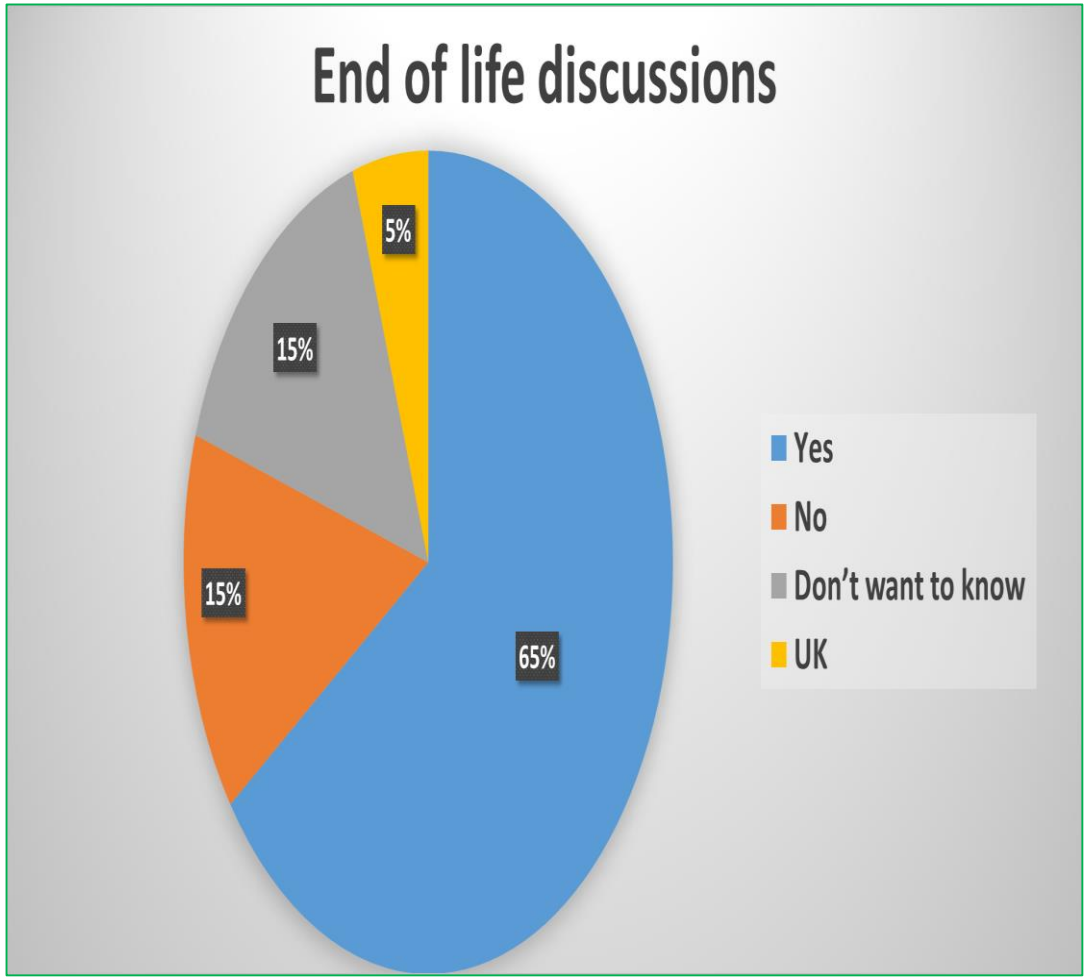
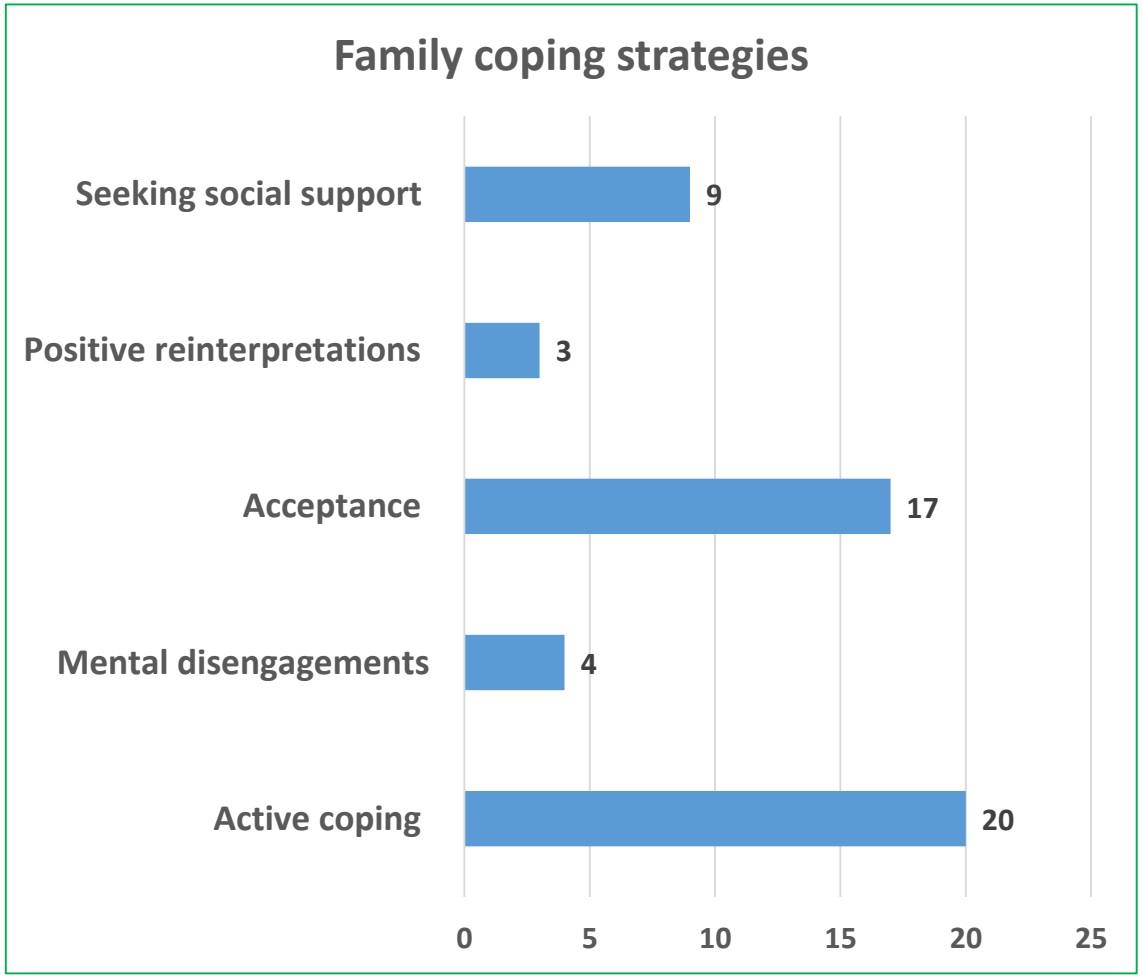




RESULTS


End of life discussions
5%
15%
15%
65%
Yes
No
Don't want to know
UK


Family coping strategies
Seeking social support
9
Positive reinterpretations
3
Acceptance
17
Mental disengagements
4
Active coping
20
0
5
10
15
20
25

DISCUSSIONS

- First study of Thalassaemia in PNG raising issues of its contribution to infant childhood mortality & morbidity which has been consistently underestimated & neglected in assessments & efforts to improve child health and survival of the children.
- 43% of the thalassaemia patients were from CP. Distribution were along the South Coasts of PNG.
- 69% diagnosed are < 1yr & majority of them were girls.
- Ratio of 1: 2 (M:F) and CFR at 5%.
- 57% were eligible for school but all not at school except one who wants to be a doctor. Prep(67%) is the highest education attained by the majority.
- NCDC residents can afford PMV fares except 3 parents (K400 per return trip for blood TF excluding other costs.)
- Thalassaemia Dx was mainly done by clinical features(54%) & known history due to no test kits.
- Post TF Hb check were not routinely done (100%) even Post TF FBC(65%). Hyper transfusion of the patients not maintained.

DISCUSSIONS CONT….

- The patients feel over protected by their family & have reduced social, physical activities
- End of life discussions was very tough but parents were understanding and cooperative of which 65% admitted to knowing their child's outcome. 15% don't want to know but leave all to God to deliberate.
- Cons clinic non attendance was mainly due to bus fare issues(58%), 52% didn't know, long distance to travel(40), work commitments(19%) & few bluntly said it was a waste of time.
- Some children like to come to the hospital because their parents buy special foods for them and lets them use the phone & are full time with them. And they do have some friends in the hospital.

SUMMARY

- Due to its chronicity patients are been branded & unintentionally over looked.
- Proper management & treatment protocols with no chelating agents had not been adhered to by all cadres of health workers needing areas of improvement.
- Thalassaemia is here to stay and with the blood transfusion treatment the age of death is elevated to < 25 years. Additionally the long family history, consanguinity with increase awareness of Thalassaemia and accessibility to health services will mean more Thal patients along the South Coasts of PNG.
- Proper and regular hyper transfusion management of the patients should improve the : Age of death. School attendances, Social activities, self esteem and other issues…
- Parents will appreciate a better blood transfusion services with introduction of chelating agents and better comprehensive care of the patients with supportive investigations & treatments with the use of other services in hospital too.
- Provision of holistic & comprehensive care for patients & families with Thalassaemia is essential including other children with Birth Defects (Born that way, R Marcus), 2019:

RECOMMENDATIONS

- Sufficient & available bloods for Day Care Management for patients to walk in & walk out
- Post transfusion Hb check/FBC before patient is sent home. Hyper transfusion!!.
- Develop a protocol card for all Thalassaemia patients and educate the parents on its use. It can be used to remind the attending health care worker of what is expected depending on the availability of services
- All Provincial Hospitals & District hospitals should have its own blood services available.
- Manage complications as well other areas like psychiatry, psychologists, nutritionists, dentists, paediatric cardiologists, haematologists etc..
- Have the health policy makers are generally aware of the global & local toll of birth defects & associated disabilities and family burdens.
- Create a Genetic Screening Services for all Birth Defects in hospitals where counselling and education of the family can be done. Collect reliable database for the country. Paediatricians to train the nurses or HEO for above.
- Health workers attending to the patients have to be compassionately encouraging & supportive.

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- (Christianson and Modell, 2004; Czeizel et al, 1993)
- Bittles, 1990; Bittles et al, 1991; Castilla et al., 1991; Christianson et al, 2000; Liascovich et al., 2001; Modell and Kuliev, 1989; Murdock, 1967; Rittler et al., 2001; WHO, 1996).

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