Diagnosis and treatment of SCD in Pemba : a joint project by Italian and Tanzanian cooperation

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Public Health Laboratory Ivo de Carneri

- WHO Collaborating Centre for NTD (Neglected Tropical Diseases)
- National TB Reference Laboratory (NRL)
- Reference Local laboratory for water analysis
- A public health service provider
- Reference Unit in Pemba for the National Health Management Information System

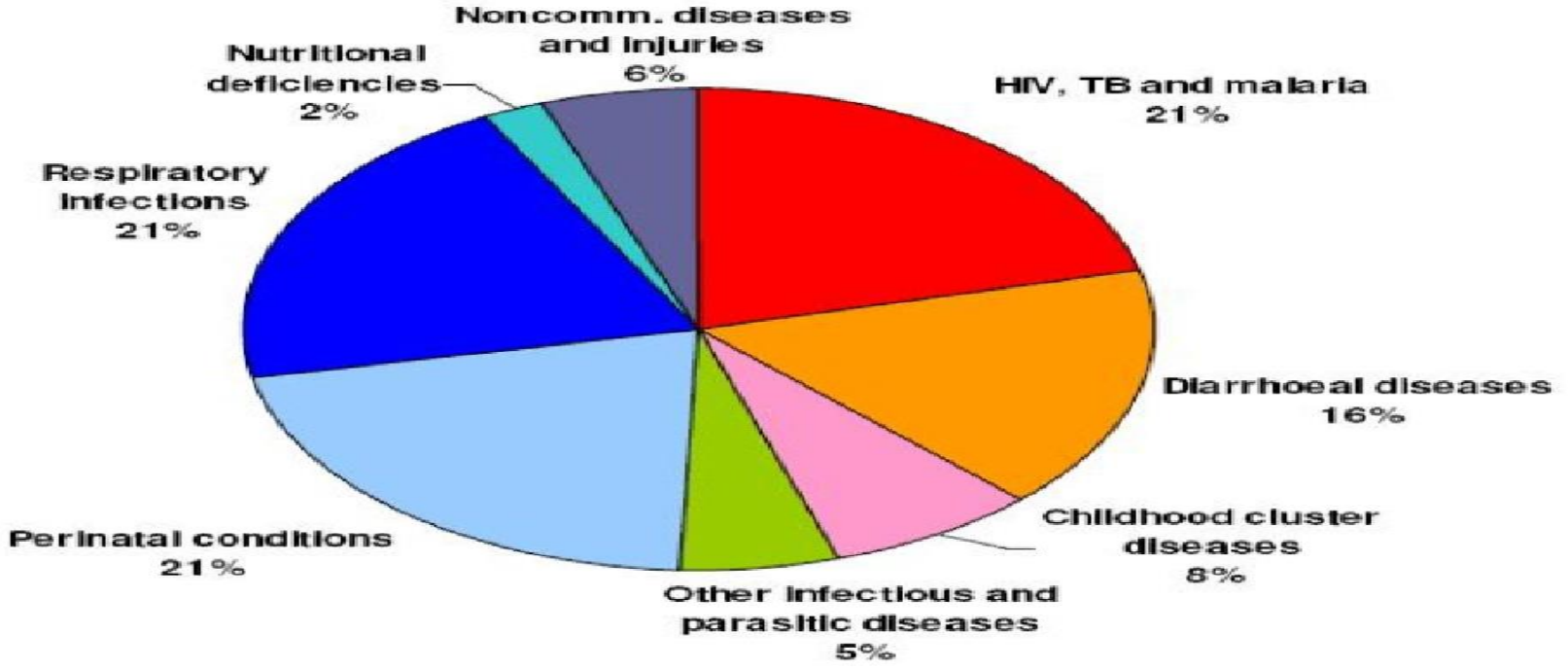
Welcome to PHL-IdC

Ivo de Carneri Foundation

Milan November 2021

SCD in Tanzania up to 2024

- **SCD birth prevalence /year**:
  6-10 per 1000 births (**around 20.000**)
- **SCD children MR/year** = 10.500 (J. Makani)
- **U5-MR for SCD = 50 to 90%**
- **U5-MR for Malaria = 10 % (WHO data)**



Source: WHO Global Burden of Disease, 2004 Update, WHO 2008

HELP3 Project : our targets

- **to decrease the SCD "U5-MR" and to improve the SCD survival and QOL, decreasing also the "social costs"**
- *to create a fruitful advocacy within National and International NGOs in favour of Tanzania*
- **to apply standard therapy first… and to cure definitively by BMT** *both childhood SCD and other blood diseases*

HELP3 «general strategy» in Pemba

1. considering the SCD a relevant social problem
2. *to involve* national health authorities
3. *to involve* Tanzanian medical and nurse personnel in a preliminary "local training" on SCD as part of social-economic improvement
4. *to approach* a correct diagnosis and treatment of childhood SCD
5. *to improve* the prognosis of SCD , both enhancing care and making it accessible to the affected children

HELP3 support in Tanzania to struggle SCD

Since 2015 collaboration,
training and echonomical support to four hospitals in Tanzania

Since 2019 at BMH (Dodoma)

*****At distance training (48teleconferences)***

***** Stages in Italy for Tanzanian nurses and medical staff of BMH (2021-2022)***

***** «Start up» of an hematologic and BMT center at Benjamin Mkapa Hospital (Dodoma) on 2023***

Direct beneficiaries up to now :

- **SCD** patients (1000)
- **Nurses/medical staff** in the hospitals (20)
- **Parents and relatives** of the SCD patients (1250)
- **Doctors** on the districts (10)

Help3 2015-2024: activities

- **SINCE 2015** : **focus on «sickle cell disease»**

➢ **four SCD "outpatients" activated in** 4 Hospitals :
TANZANIA : BMC -Mwanza, **St.Gemma H**-Dodoma , **Mnazi Mmoja H** -Zanzibar
UGANDA : **Lacor hospital**

➢ **Donation of Hydroxyurea** for the treatment **of 1000 SCD children**

➢ **Donation of "data base" SCD oriented**

➢ **Donation of "Electrophoresis of Hb" instruments to 3 Tanzanian Hospitals**

➢ **Italian hematologysts available every day for consultation** (4)

- **SINCE 2019 : focus on «Bone marrow transplantation»**

➢ ***Collaboration with BMH (Dodoma) toward the Start up of Haematologic and BMT Unit for the diagnosis and treatment of SCD and other hematologic diseases***

➢ ***First 8 BMTs successfully performed at BMH (Dodoma) in SCD children***

➢ ***Twenty Italian professionals involved at the «start up» of BMT Unit 2023-2024***

Help3 «specific strategy» in PEMBA to struggle childhood SCD

1. *Supporting an "educational project" involving local professionals , schools and parents of SCD in Pemba , i.e distributing "booklets" in English or local language Swahili*

2. *Providing for free the Lab instrument* ("point of care" Gazzelle) for the diagnosis of SCD and *promoting "internship"* of 1 Lab technician

3. *Providing for free the hydroxyurea treatment* at least for the first **50** children recognized as affected with SCD

4. *involving Pemba professionals* on the management of SCD treatment

5. *Offering an "on Line Data Base"* , for a very simple "collection of data" in the setting of :

a) a better surveillance of patients follow up

b) a fruitful system of "*at distance/on line suggestions or second opinions "*

Help3 and 5 years interactivity at St.G.H , Dodoma

Involving the personnel

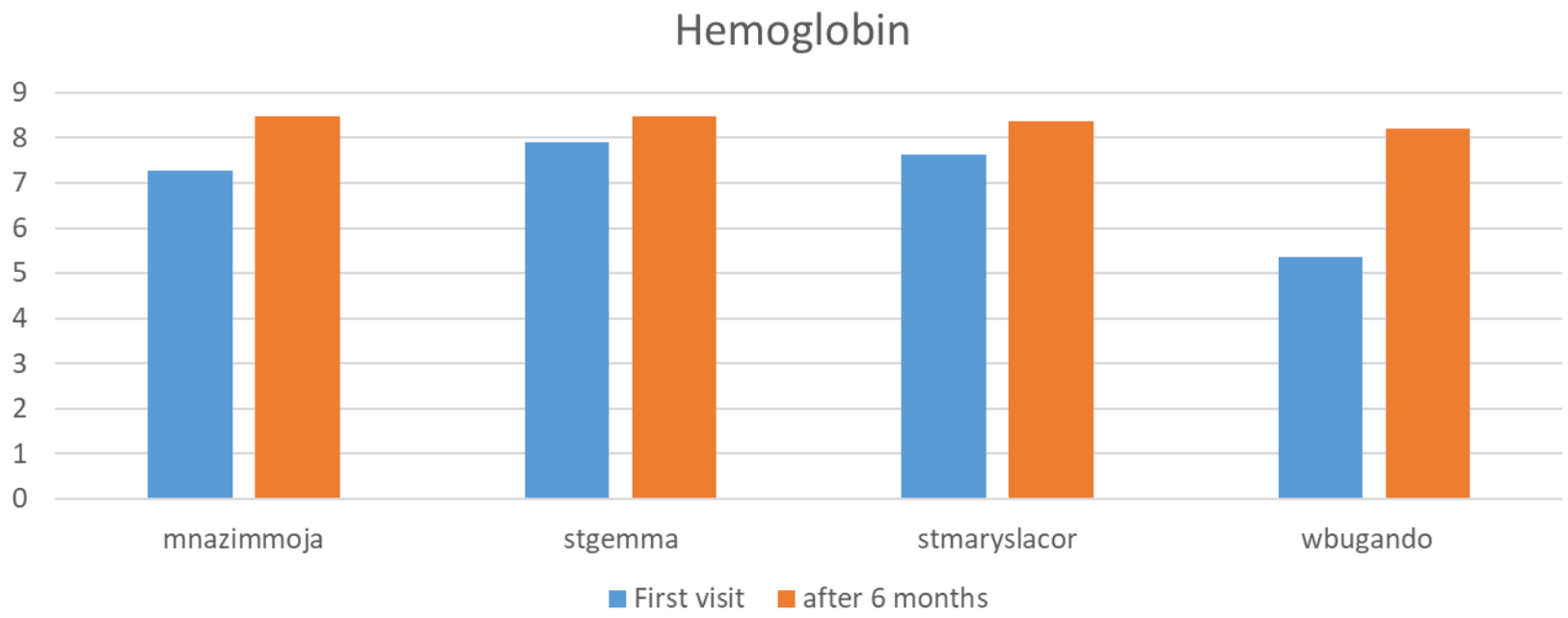
Sickle cell clinic:

450 SCD treated children

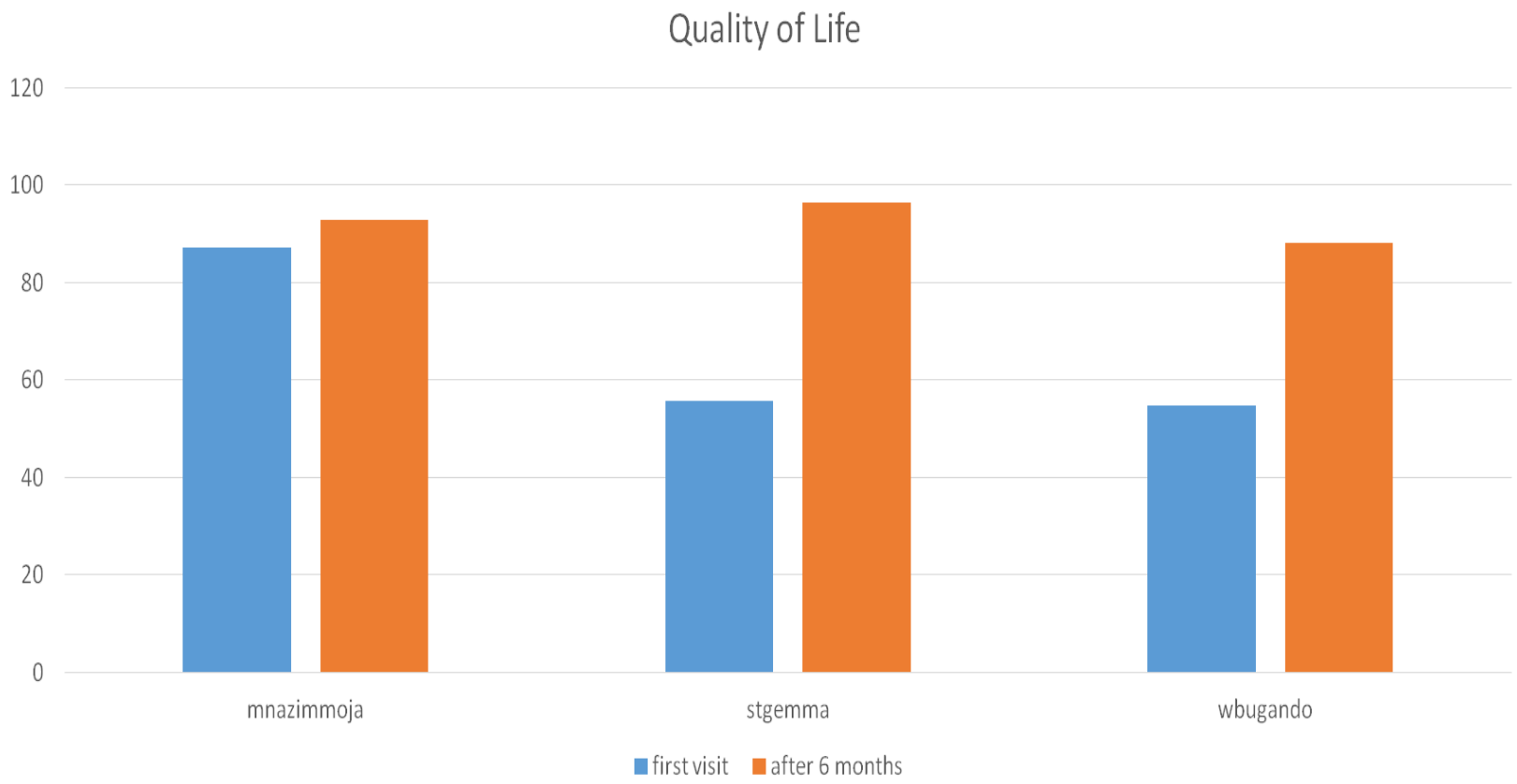
Help3 and 3 years interactivity at MMH

MNAZI MMOJA HOSPITAL (ZZ) :
200 SCD treated children

Median Hb value in 249 SCD children pre and post HU



Median QOL in 196 SCD children pre and post HU



Hydroxyurea with dose escalation for primary stroke reduction in children with SCD in Tanzania : an open-label, phase 2 trial

Emmanuela E Ambrose [1], Teresa S Latham [2], Primrose Songoro [3], Mwesige Charles [4], Adam C Lane [2], Susan E Stuber [5], Abel N Makubi [6], Russell E Ware [7] et a. LANCET Hematol 2024

- Children with SCD in Tanzania have a high baseline stroke risk
- *Chronic transfusions* **reduce** stroke risk in children with sickle cell anaemia but is not feasible in low-resource settings.
- *Hydroxyurea* is an alternative treatment to decrease stroke risk.

- *Hydroxyurea* at the maximum tolerated dose significantly lowers transcranial Doppler velocities and reduces primary stroke risk.
- *Transcranial Doppler screening plus hydroxyurea* at the maximum tolerated dose is an effective stroke prevention strategy, supporting wider hydroxyurea access for patients with SCD across sub-Saharan Africa

WHY BMT for "SCD" in childhood ?

- **is the only "curative treatment "** for **SCD** since 20 years in "high income countries" ***with 95% of "cure rate"*** and no *recurrence of clinical "vaso-occlusive crisis"* and no new ischemic lesions after successfull transplant
- **High "quality of life" after BMT**

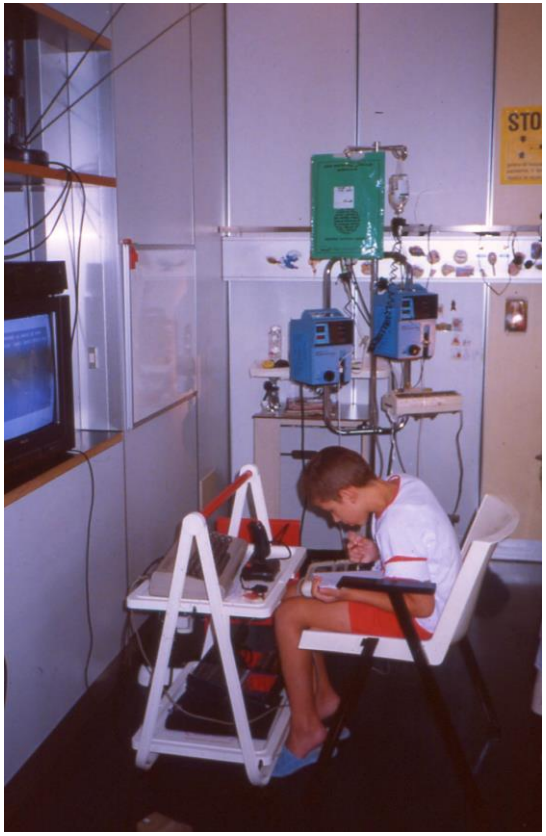
WHY BMT for malignant diseases in childhood ?

- ***High "cure rate" (60-70%) in patients with "resistant diseases" after standard chemotherapy***
- **BMT success in a large series of patients all over the world since last 2 decades**
- **BMT , Innovative treatment , is feasible today also in "developing countries" including "Subsaharan area"**

«BMT» at Benjamin Hospital: 2023-2024

Bone marrow harvesting

Bone marrow infusion



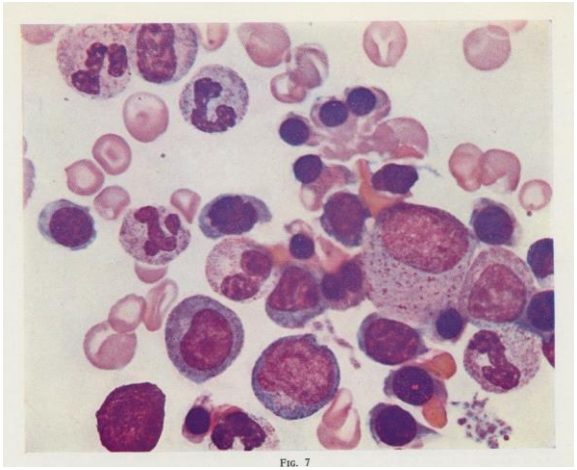


FIG. 7

CONCLUSION 1 : there is a strong need for launching «SCD project» in Pemba island

CONCLUSION 2

children must have the «right to health» all over the world

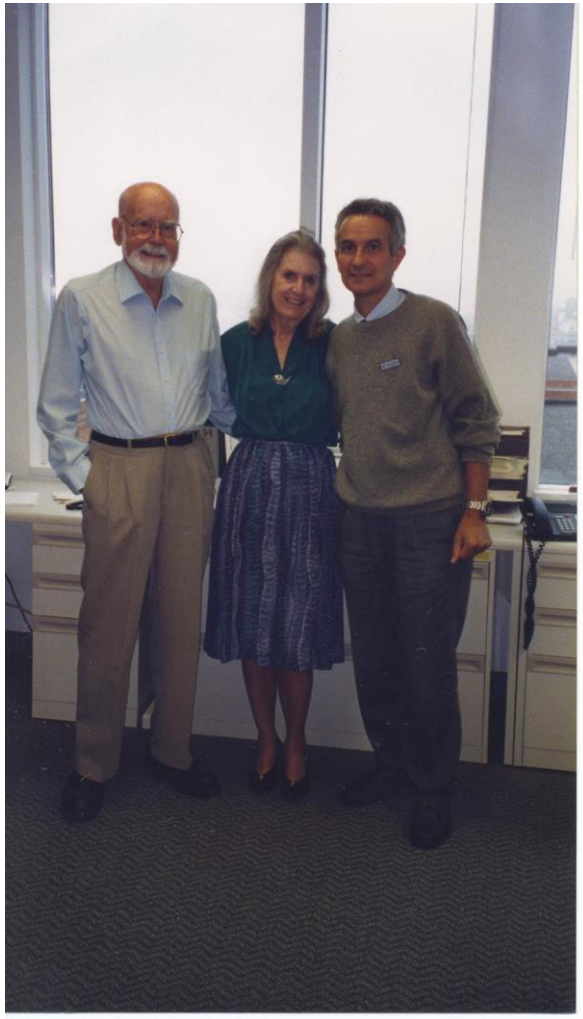
A big thanks to

Donnall Thomas…

BMT pioneer,

Nobel Prize on 1990

and a great "life coach"



CONCLUSION 2

The development of curative treatment for children with hematologic diseases is a benchmark for medical progress and such treatment *must not be sequestered within the borders of few countries*

Raul C. Ribeiro and Ching Hon Pui

NEJM may 2005

BMT BASAL REQUIREMENTS

OUTPATIENT FACILITIES (with air conditioning)

STAFF : 2 doctors + 4 Medical doctors ; 8 (10) nurses;1 chief of nurses

BASAL STRUCTURE :

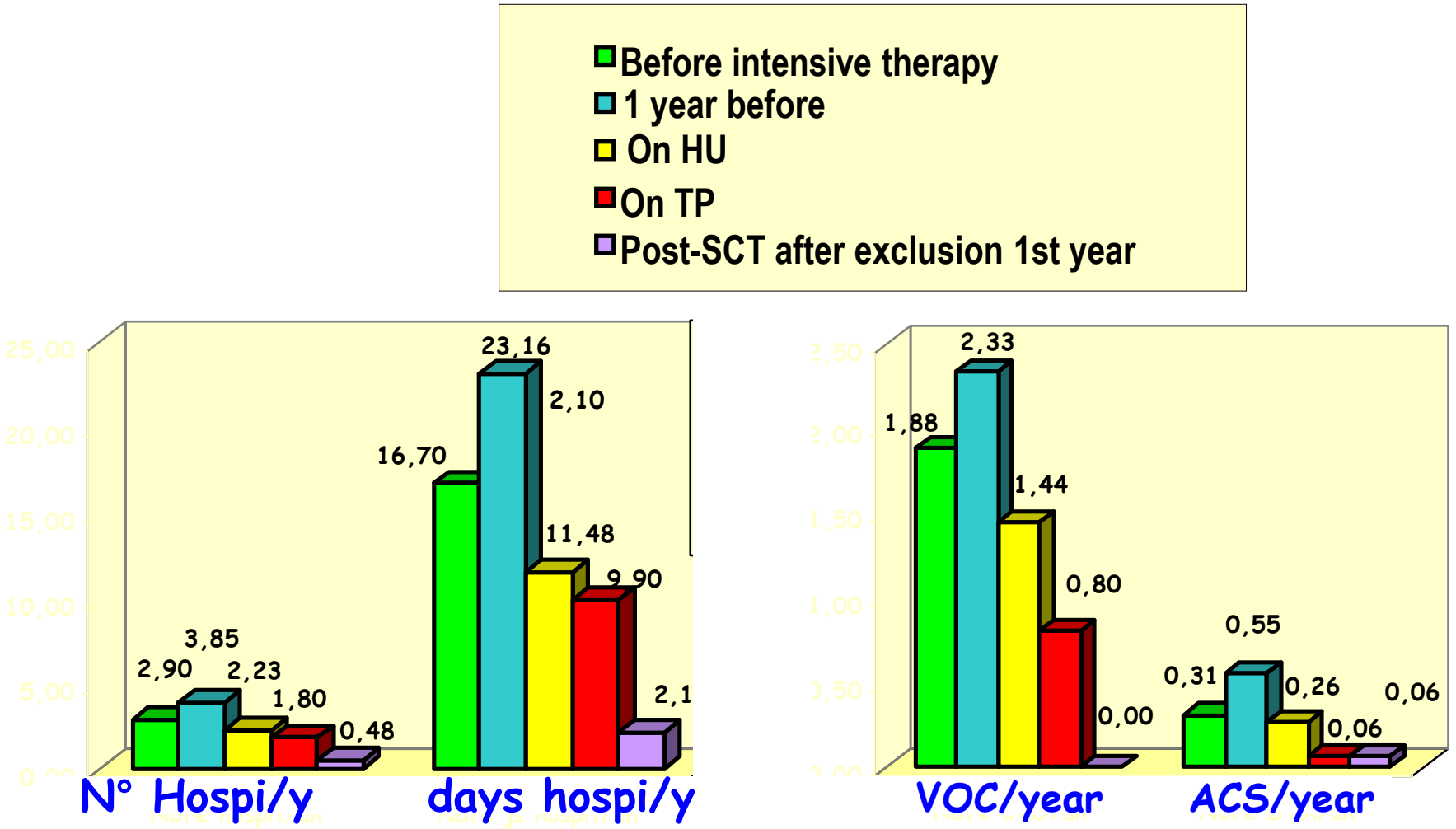
- **large common room with 10 beds for transfusions, infusions + 4 bathrooms**
- **2 rooms for invasive and non-invasive procedures (BM aspiration, lumbar puncture……..**
- **1 *Nurses station* for preparation of infusions and any kind of procedure (+ Internet/Intranet)**
- **1 *Doctors station* with Internet/Intranet**

OUTPATIENT FACILITIES (with air conditioning

SUPPORT AEREA :

- **One protective cabinet**
- **Storage room (for drugs, (scales, monitors…….)**
- **1 "Play/rest room" for patients and/or parents**
- **1 rest room and 2 bathrooms for the personnel and parents**
- **1 room for "social worker and/or psycologyst**

Comparative effects of TP, HU and BMT on frequency of Hosp, VOC, ACS in 111 SCD-patients



Arnaud et al CHI-Creteil, ASH