

ABHH Webinar, 29 DE JULHO, 19H
O IMPACTO DA COVID-19 NAS ANEMIAS

Nas pessoas com talassemia

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COVID-19 and β -THALASSEMIA

- Current experience in thalassemia patients?
- Are thalassemia patients at higher risk?
- Implications to management of β -thalassemia patients?

COVID-19 and β -THALASSEMIA

- Current experience in thalassemia patients?

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covid-19 AND thalassemia

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1

A Case of **COVID-19** in a
Thalassemia and Glucose-6-Phosphate Dehydrogenase Deficiency.

Sasi S, et al. *Am J Case Rep.* 2020. PMID: 32697769

CASE REPORT We report the case of a young man with no known co-morbidities, who came with

20 publications up to July 29, 2020

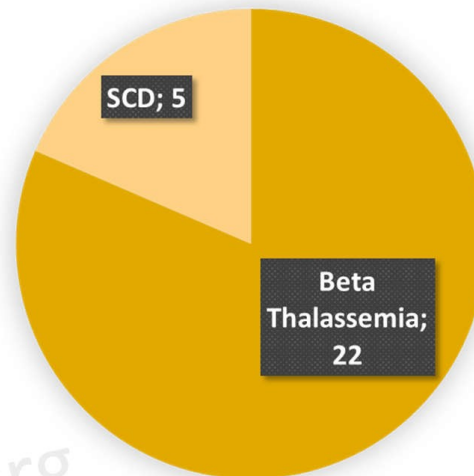
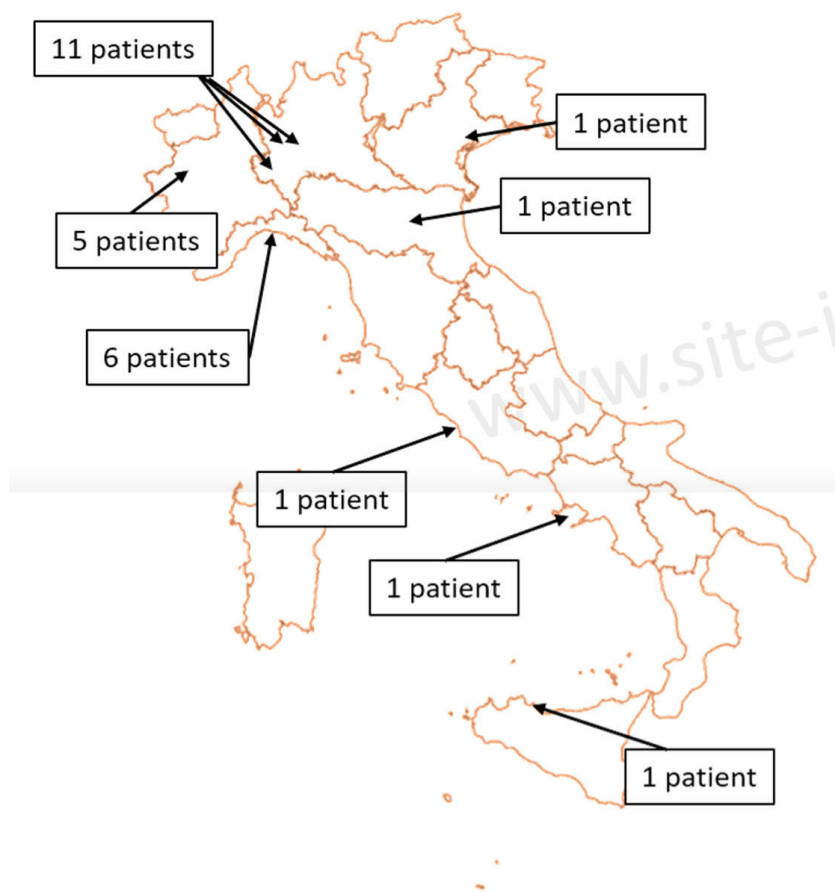
- 2 clinical studies
- 2 case reports
- 16 opinion papers

➤ [Am J Hematol.](#) 2020 Apr 20;10.1002/ajh.25840. doi: 10.1002/ajh.25840.
Online ahead of print.

SARS-CoV-2 Infection in Beta Thalassemia: Preliminary Data From the Italian Experience

Irene Motta^{1 2}, Margherita Migone De Amicis², Valeria M Pinto³, Manuela Balocco³, Filomena Longo⁴, Federico Bonetti⁵, Barbara Gianesin⁶, Giovanna Graziadei², Maria D Cappellini¹, Lucia De Franceschi⁷, Antonio Piga⁴, Gian L Forni³

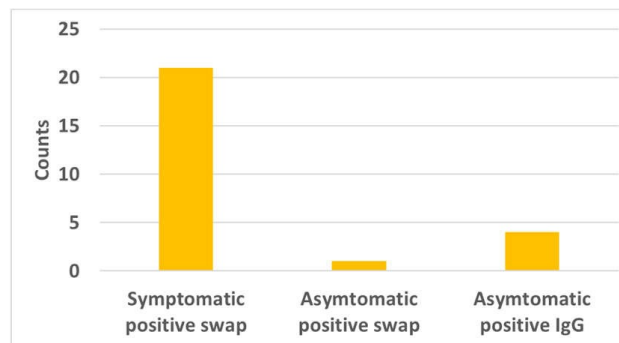
Regional distribution



27 patients: 22 β -thalassemia, 5 SCD

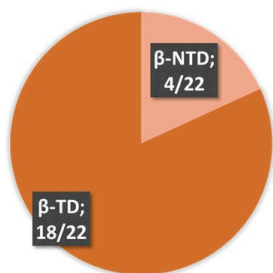
52% males

Mean age (SD): 41 years (13)



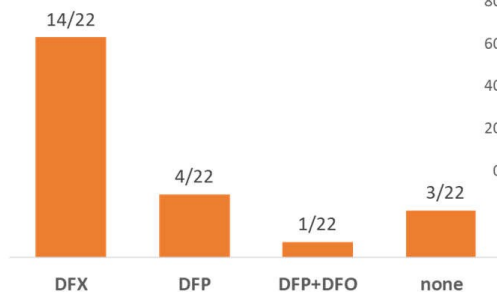
THALASSEMIA PATIENTS

Characteristics of the patients

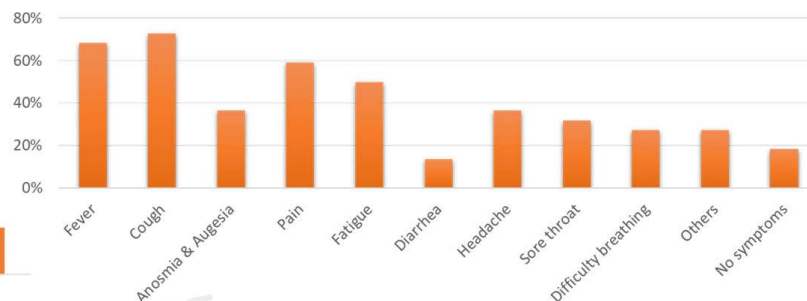


#22 patients β -thalassemia
55% males,
Mean age (SD) : 43 years (12)

Iron chelation therapy

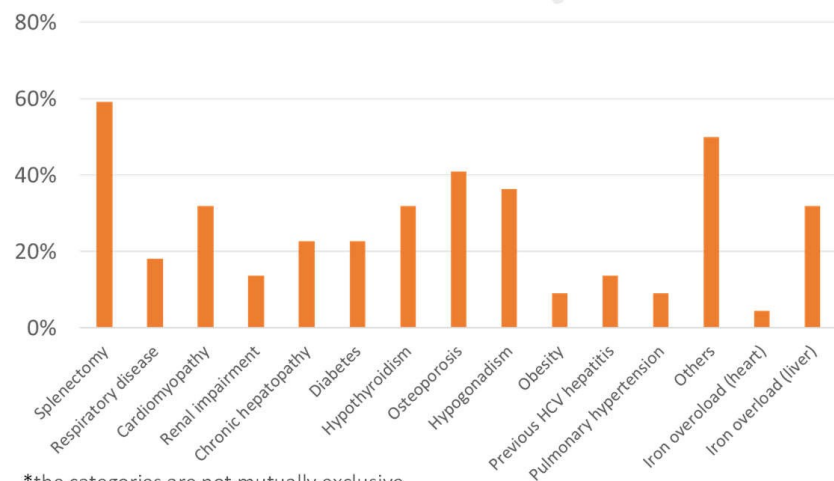


Symptoms*



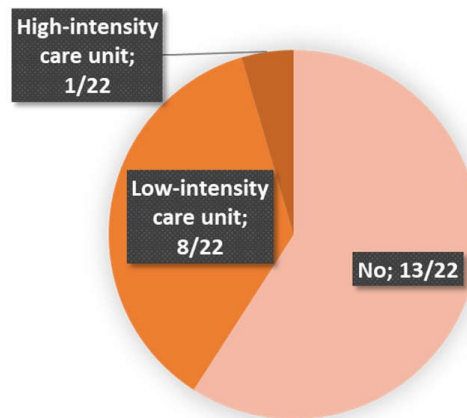
*the categories are not mutually exclusive

Comorbidities*



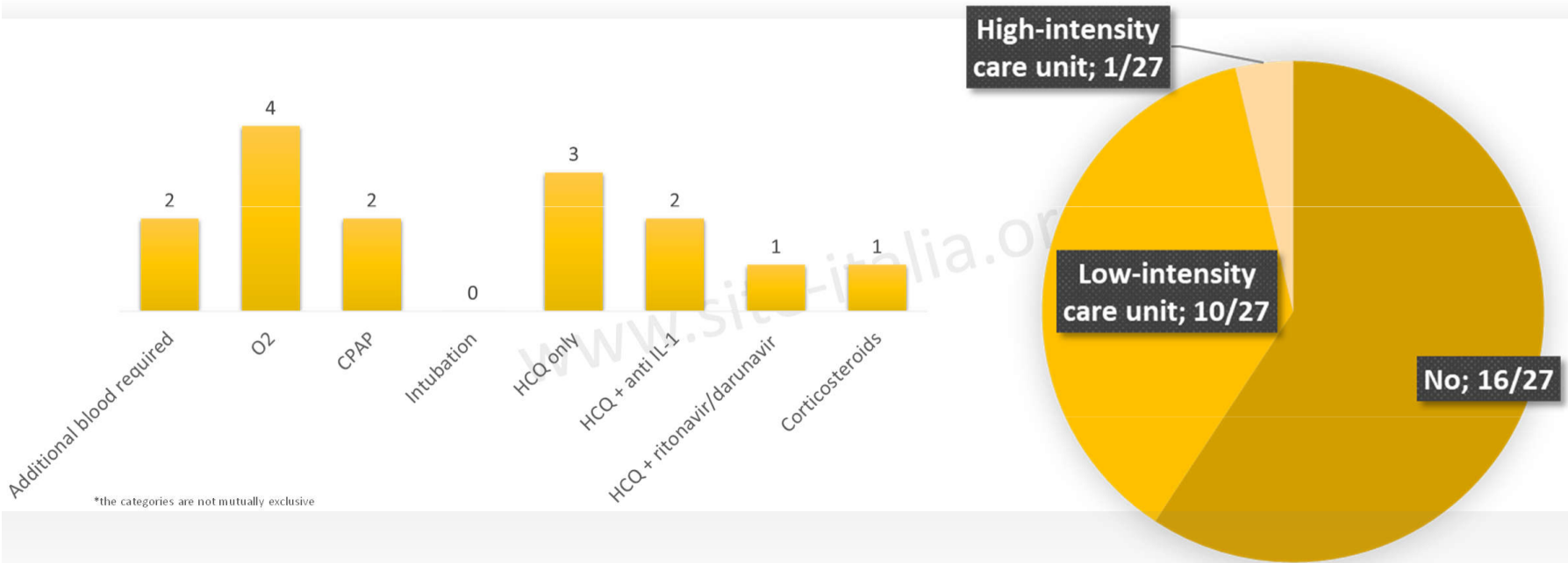
*the categories are not mutually exclusive

Hospitalization



HEMOGLOBINOPATHIES all pts

Sars2-Covid19 Specific therapies*



Outcome

100% recovered

**Clinical Course of hospitalized pts:
18 days (7)**

COVID-19 and β -THALASSEMIA

Italian study

- **24 cases**
(20 TDT and 4 NTDT)
- **Prevalence**
3,5/1.000 (3,9/1.000 in general population)
- **Exposure**
Professional risk or family contact in most
- **Severity**
92% mild to moderate symptoms
- **Blood requirement**
No increase
- **Mortality**
0%

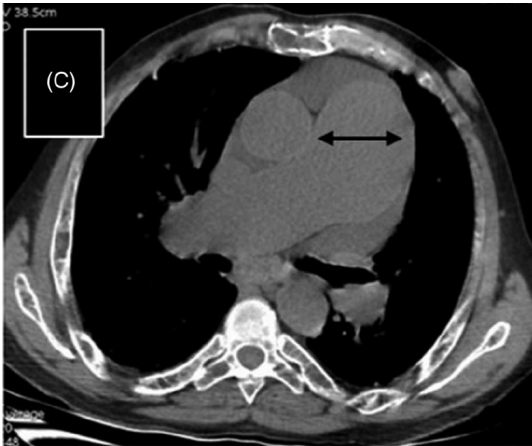
SHORT COMMUNICATION



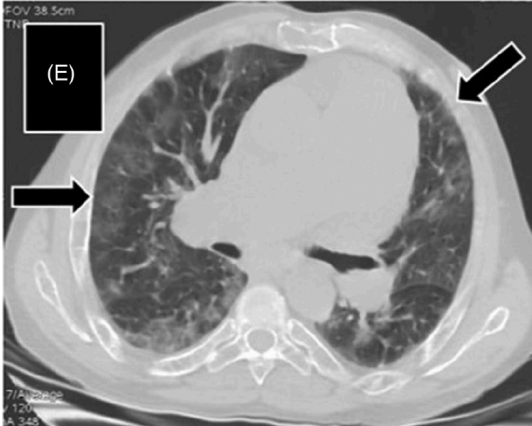
COVID-19 in a Patient with β -Thalassemia Major and Severe Pulmonary Arterial Hypertension

March 2020
COVID-19 Disease

Chest CT with enlarged main pulmonary artery

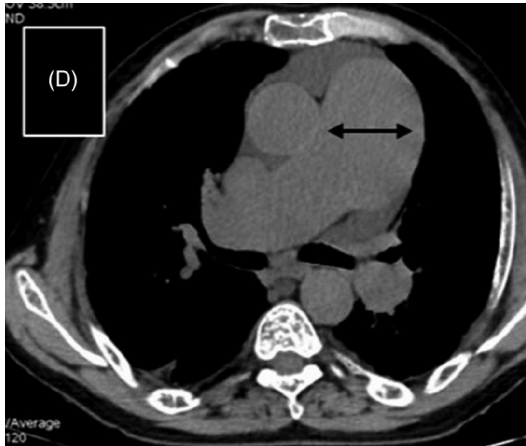


Bilateral ground glass opacities (black arrow)

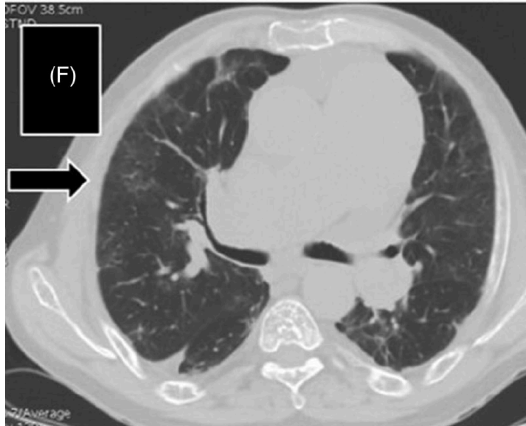


April 2020
Recovery

Chest CT with unchanged PAH



Almost complete resolution (black arrow)





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> [Am J Case Rep.](#) 2020 Jul 22;21:e925788. doi: 10.12659/AJCR.925788.

A Case of COVID-19 in a Patient with Asymptomatic Hemoglobin D Thalassemia and Glucose-6-Phosphate Dehydrogenase Deficiency

[Sreethish Sasi](#)¹, [Mohamed A Yassin](#)², [Arun Prabhakaran Nair](#)³, [Muna S Al Maslamani](#)³

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A Case of COVID-19 in a Patient with Asymptomatic Hemoglobin D Thalassemia and Glucose-6-Phosphate Dehydrogenase Deficiency

Authors' Contribution:

Study Design A

Data Collection B

Statistical Analysis C

Data Interpretation D

Manuscript Preparation E

Literature Search F

Funds Collection G

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Conflict of interest: None declared

Source of support: Qatar National Library

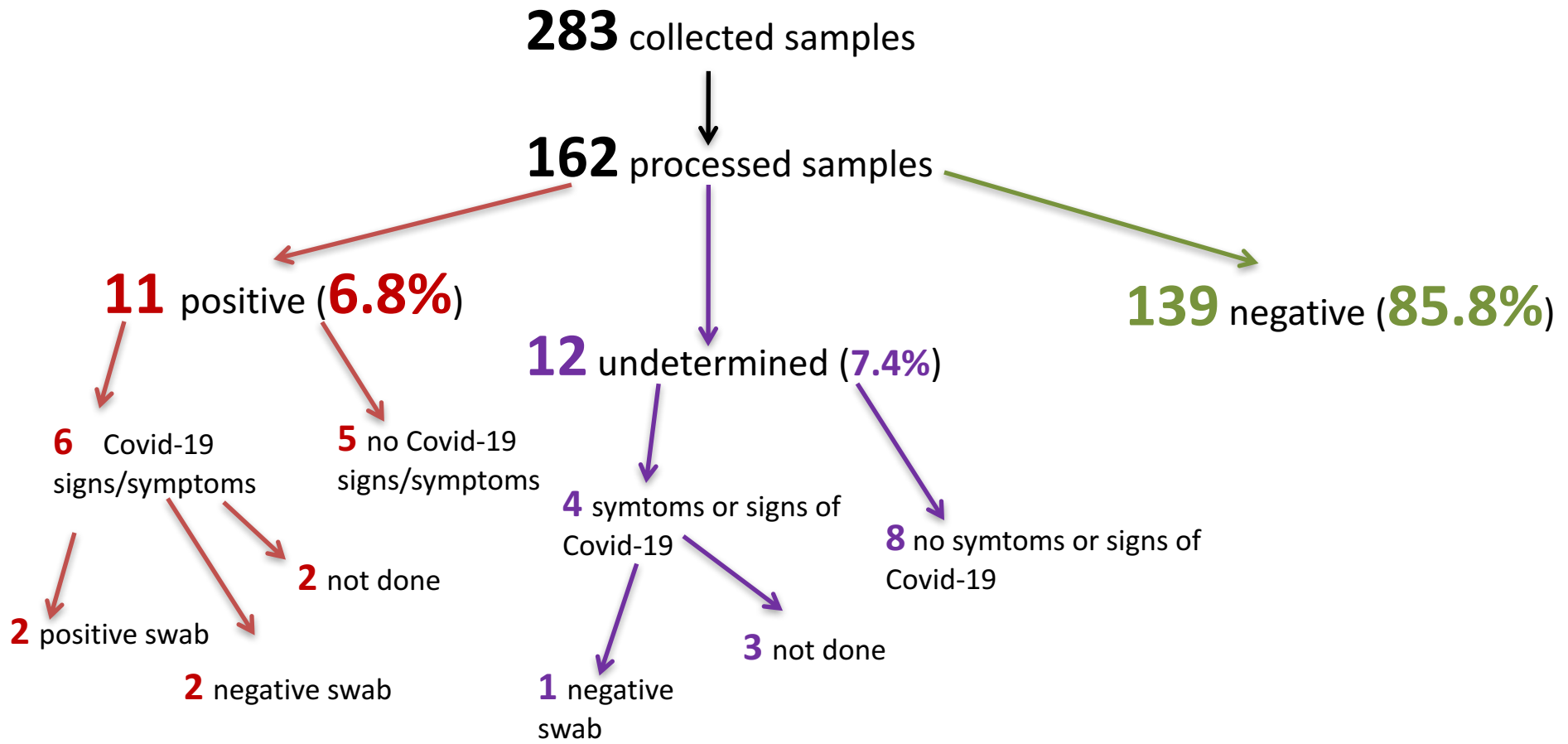
Patient: Gender, 26-year-old

Final Diagnosis: COVID-19

Symptoms: Cough • fever

Hb Covid-19 Orbassano: Prevalence of SARS-CoV-2 infection in patients with thalassemia, other hemoglobinopathies and rare anemias

Preliminary results (20 July 2020):



Hb Covid-19 Torino: Prevalence of SARS-CoV-2 infection in patients with thalassemia, other hemoglobinopathies and rare anemias

Piedmont:

○ Population	4.365.000
○ Covid-19 cases	32.000
○ Prevalence	7.3 ‰

Torino Hb-pathies Centre:

○ Population	633
○ Covid-19 <u>preliminary</u> cases	11
○ <u>Preliminary</u> prevalence	17 ‰

➤ Br J Haematol. 2020 Jun 2. doi: 10.1111/bjh.16911. Online ahead of print.

Prevalence and Mortality Due to Outbreak of Novel Coronavirus Disease (COVID-19) in β -Thalassemias: The Nationwide Iranian Experience

Mehran Karimi¹, Sezaneh Haghpanah¹, Azita Azarkeivan², Zohreh Zahedi¹, Tahereh Zarei¹, Maryam Akhavan Tavakoli³, Asghar Bazrafshan¹, Afshan Shirkavand⁴, Vincezo De Sanctis⁵

COVID-19 and β -THALASSEMIA

Iranian study

- **23 cases**
(18 TDT and 5 NTDT)
- **Prevalence**
0,8/1.000 (2,2/1.000 in general population)
- **Exposure**
?
- **Severity**
74% mild to moderate symptoms
- **Mortality**
26% (2 TDT and 4 NTDT; most with clinical complications)

COVID-19 and β -THALASSEMIA

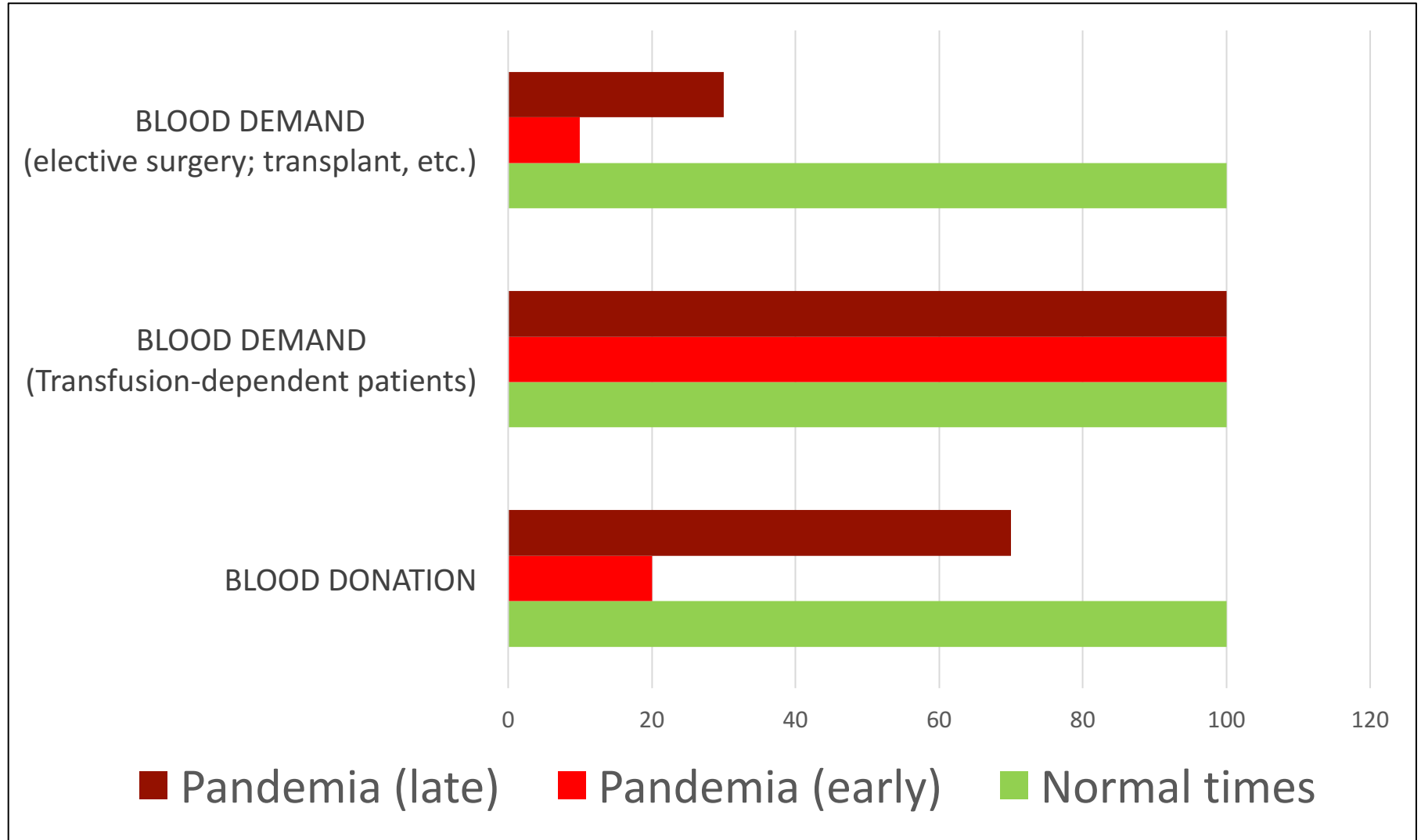
- Current experience in thalassemia patients?
- Are thalassemia patients at higher risk?

COVID-19 and β -THALASSEMIA

Are thalassemia patients at higher risk?

- **Blood**
 - No data suggesting a risk of transfusion transmission
 - No evidence of non-AB blood groups protection
 - No serious shortage of blood

COVID-19 and Blood balance



COVID-19 and Blood balance

	Considerations	Possible actions
Donor recruitment ^{34,46-48}	Donors tend to respond well to public appeals in situations of perceived exceptional need (eg, September 11 attacks and mass shooting events); a large influx of donors is to be expected, at least initially; donors are more tolerant to longer waiting times; platelet donations require close attention because of their short shelf life; some donors might be prevented from donating because of stay-at-home orders (eg, older, reliable donors)	Encourage appointments but discourage walk-ins; track donor characteristics (first-time donor vs repeat donor, as well as age, sex, etc); reinforce platelet apheresis donation programmes; consider increasing reliance on whole blood-derived platelets; more forcefully target first-time and reactivated donors for future donations
Donor eligibility ^{40,49-53}	Some donor-selection criteria could be relaxed without any meaningful effect on donor or product safety (for examples, see possible actions column); this approach can only be justified if supply cannot meet demand and changes need to be planned in advance because of their complexity (eg, regulatory aspects, IT system changes, and training of personnel); consideration should be given on the acceptability of reinstating pre-pandemic criteria after the pandemic is over (easier to explain to donors for some measures [eg, Haemoglobin concentrations] than others, such as reinstating permanent deferrals for variant Creutzfeldt-Jakob disease); some procedures can also be interrupted to increase compliance with public health recommendations, including social distancing; the COVID-19 situation might exacerbate criticisms over deferral policies for men who have sex with men, although shortening the deferral period will likely yield few additional donors	Discussions could be held with regulatory authorities regarding mechanisms for urgent implementation and expedited reviews; some procedures and criteria regarding donor safety could be considered for relaxation (eg, salty snacks on blood drives before and during donation, heart rate and blood pressure measurements, interdonation intervals, haemoglobin thresholds, and age restrictions); some procedures and criteria regarding recipient safety could be considered for relaxation (eg, deferral period for travel in a malaria-risk area; deferral period for tattoos, piercings, and needle-stick injuries; deferrals for men who have sex with men; and deferrals for variant Creutzfeldt-Jakob disease risk)
Blood drive planning ³⁶	Decreasing and increasing demand; suitability of donation sites; public health recommendations and governmental communications regarding confinement; public appeals for donation; staff availability	Adjust the number and size of upcoming blood drives; review physical distancing requirements when choosing locations for mobile blood drives; consider expanding collections on fixed sites; work with public health advisors and government communicators to emphasise the importance of blood donation as a reason for travel; work with health authorities to coordinate public appeals for donation, if and when appropriate
Inventory management ⁵⁴	Demand is hard to predict and might vary in different phases of the pandemic	Keep close contact with hospital customers, including regular updates of inventory; track system-wide inventory closely; monitor activities requiring increased blood use (eg, elective surgery and transplantation)
Protection of staff and donors ^{36,47,55-58}	Use of personal protective equipment for donors, staff, and volunteers; practice physical distancing; monitor COVID-19 illness among staff and donors; message donors before arrival on the blood drive regarding wellness; prescreening for COVID-19 signs and symptoms	Align practices with public health recommendations; review availability of personal protective equipment; implement a communication plan for occupational risk; disseminate guidelines for COVID-19 signs and symptoms among personnel, donors, and volunteers (quarantine and testing, etc); consider screening donors, personnel, and volunteers for symptoms and elevated temperature before entering facilities and donation sites
Availability of personnel ^{35,40,53,60}	Effect of COVID-19 on staff: illness, quarantine, and fear of disease	Prepare contingency plans for staff replacement (eg, reassignment and training of other non-essential staff); communicate clear supportive policies for sick leave; encourage staff to self-report illness or concerns; offer and strengthen psychological support for personnel
Plasma for fractionation ^{61,62}	The effect on supply of plasma for fractionation is uncertain, including the supply of immunoglobulins; blood providers might temporarily decrease their source plasma donation programmes to shift their capacity to whole blood donations	Efforts should be made to maintain or increase source plasma donations in the context of the pandemic; reconsider the need for certain procedures and criteria in donor screening, such as the annual physical exam; blood providers could take advantage of the influx of new and repeated donors to increase collections of source plasma collections
Product safety ^{38-45,63}	To date, there is no evidence of SARS-CoV-2 transmission by transfusion; some infected people appear to have detectable RNA in their blood, even when they do not have severe symptoms; RNA has been found in a few blood donors, but the concentrations are low, and the results might represent false positives; RNA in blood does not necessarily represent infectious viral particles; the South Korean lookback study ³⁹ found no evidence of transmission	Do additional studies to establish the presence of virus in blood donors; do lookbacks and tracebacks when appropriate; reinforce postdonation information protocols; evaluate the availability and appropriateness of blood screening tests for donors; communicate risk assessments to relevant stakeholders; eligibility criteria should be applied to reduce the risk of collecting blood from infected donors; deferral periods should be applied for confirmed or suspected cases, for travel in countries or regions at high risk, and for exposure to confirmed cases (also important for safety of staff and other donors)

SARS-CoV-2=severe acute respiratory syndrome coronavirus 2.

Table 1: Donor and donation factors to consider for maintaining an adequate supply of blood during the COVID-19 pandemic

COVID-19 and β -THALASSEMIA

Are thalassemia patients at higher risk?

- **Blood**
 - No data suggesting a risk of transfusion transmission
 - No evidence of non-AB blood groups protection
 - No serious shortage of blood
- **Splenectomy**
 - No evidence of enhanced risk
- **Iron**
 - No evidence of association of iron overload and clinical severity
 - Iron chelation tested in vitro not in vivo

COVID-19 and β -THALASSEMIA

- Current experience in thalassemia patients?
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Algorithm for assessment of patients with symptoms suggestive of COVID-19 infection

Triage of haemoglobinopathy and rare anaemia patients presenting to hospital with fever, cough, myalgia, or other symptoms* suggestive of COVID-19 infection



Assessment should include evaluation of possible bacterial sepsis; blood count and biochemistry including CRP, blood culture, O₂ saturation, NPA for respiratory viruses + COVID-19 screening**

**Loss of smell (anosmia) or taste (hypo/ageusia) is reported in a significant proportion of cases*

***If not admitted, patients should be contacted with their COVID-19 result and advice on self-isolation*



Sickle cell disease

If no acute pain crisis, signs of sepsis or neutropenia due to hydroxycarbamide, T < 38°C, CRP < 100, O₂ sat ≥ 95% or normal PaO₂ on ABG on room air: monitor at home +/- oral antibiotics with instructions to self-isolate and contact centre for urgent advice if not improving or if at any time if short of breath.

Thalassaemia and rare anaemias

All patients should be discussed with a senior clinician in the specialist team before reaching a decision to monitor at home. If febrile, interrupt chelation until FBC and renal function can be checked. Patient may need continuous IV chelation if heart failure or dysrhythmia develop.

Sickle cell disease

Admit to hospital if any of the following: 1) Acute pain crisis requiring regular parenteral analgesia 2) T ≥ 38°C or signs of sepsis 3) O₂ sat ≤ 94% or hypoxia on ABG on room air 4) RR > 20 bpm or equivalent age adjusted 5) Neutropenia

Thalassaemia and rare anaemias

All patients should be discussed with a senior clinician in the specialist team.

CORRESPONDENCE

Care of patients with hemoglobin disorders during the COVID-19 pandemic: An overview of recommendations

To the Editor:

The outbreak of Coronavirus Disease 2019 (COVID-19) caused by the novel severe acute respiratory syndrome coronavirus 2 (SARS-CoV-2) has led to a global health emergency.¹ Compared to the general population, patients with hemoglobin disorders such as sickle cell disease (SCD) or thalassemia are expected to be more severely affected by COVID-19 due to their preexisting chronic morbidities.² The Centers for Disease Control and Prevention does not report any specific indications for patients with hemoglobinopathies. However, it can be hypothesized that the rapid spread of the virus may render these patients fragile when fighting the infection.

SCD, a hematological condition with functional asplenia, puts patients at a greater risk to develop acute pulmonary complications, including viral infections.² A study by Hussain et al reported four SCD

changes. Exchange transfusions should be initiated in case of progressively worsening hypoxemia and clinical deterioration. Blood products shortage is anticipated during the pandemic, so pre-established transfusion thresholds should be adjusted to include mainly patients with severe anemia or with complications, namely ACS or stroke. There currently exists no evidence that being on hydroxyurea would increase SARS-CoV-2 infection risk. However, it is advisable to avoid the routine use or increasing doses of hydroxyurea to reduce the need for repeated phlebotomy and hospital visits.^{2,6} In areas where severe blood shortages are expected due to the pandemic, a low dose of hydroxyurea is recommended in all pediatric patients with sickle cell anemia, who receive regular blood transfusion therapy for primary or secondary stroke prevention.⁷ In the absence of regular blood transfusion therapy, hydroxyurea treatment will also

COVID-19 and β -THALASSEMIA

Management

- **Keep**
 - Standard transfusion regimen
 - Standard iron chelation regimen
- **Consider**
 - Low dose corticosteroids (adrenal insufficiency)
 - Prophylactic anticoagulation (hypercoagulable state)
 - Antibiotics (overwhelming sepsis in splenectomy)
- **Defer**
 - Stem cell transplant or Gene therapy programs
- **Balance protocol vs. clinical needs**
 - Patients running a clinical trial

COVID-19 and β -THALASSEMIA

- Current experience in thalassemia patients?
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- Implications to management of β -thalassemia patients?