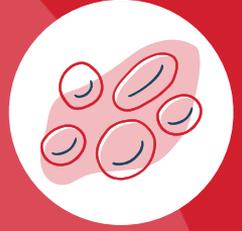


# 18<sup>e</sup> Journée de la filière MCGRE



## Intérêt de l'exercice physique dans la $\beta$ -thalassémie transfusion-dépendante

28 juin 2024

Salle Adicare

Institut de Cardiologie  
Groupe Pitié-Salpêtrière  
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Mathilde Noguier  
Merci !

MCGRE  
FILIERE SANTE MALADIES RARES



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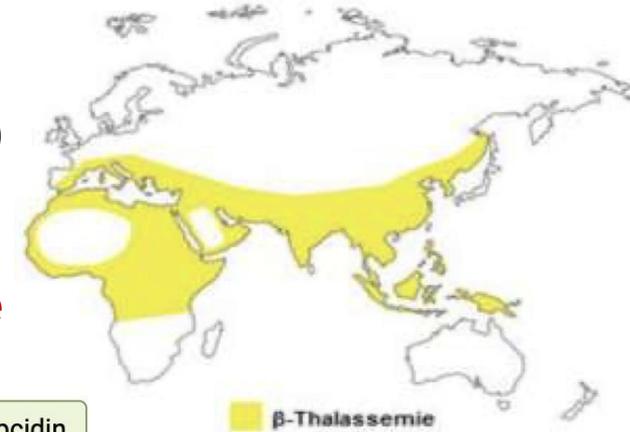


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de France

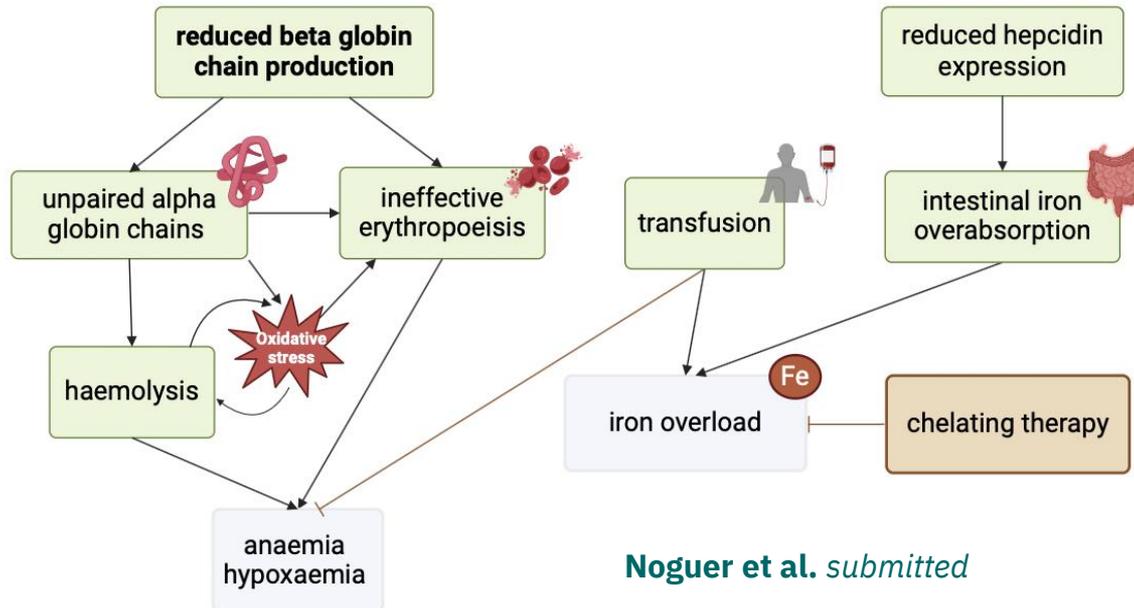


# 1- Rappels sur la physiopathologie

- Mutations génétiques (chromosome 11, gène HBB)
- Hémoglobinopathie autosomique récessive



## Faible production de chaîne beta de l'hémoglobine



## 2- Rôle délétère de la surcharge en fer sur les organes impliqués dans l'aptitude physique

### 1. Poumons

Chan et al. *Sci Rep* 2023

### 2. Cœur

Detterich et al. *Am J Hematol* 2012

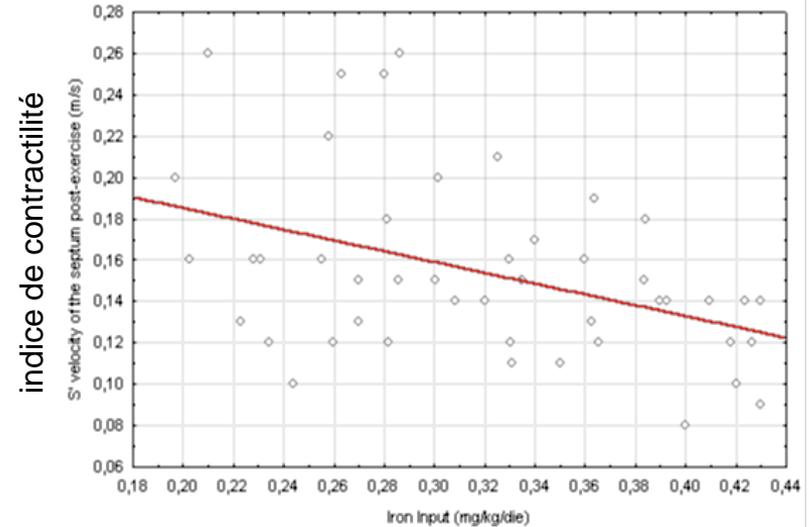
### 3. Os

Rossi et al. *Haematologica* 2014

### 4. Muscle et Nerfs

Nemtás et al. *Ann Hematol* 2018

Barbero et al. *Mediterr J Hematol Infect Dis* 2012



relation between iron input during the previous year (2010) and S' waves measured by TDI on the basal segment of the septum after exercise ( $r = -0,413$   $p = 0,004$ ).

### 3- Rôle délétère de la chélation

Si la chélation diminue les risques d'atteintes d'organes (cardiopathie) et augmente l'état de santé, la qualité de vie, la survie...

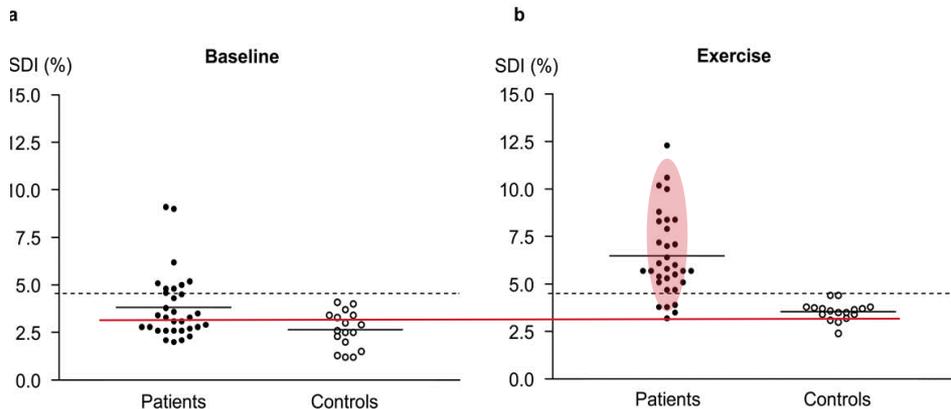
**Elle peut aussi avoir des effets délétères, notamment sur le muscle...**

| Variables                         | Overall sarcopenia |         |                    |         | Severe sarcopenia  |         |                    |         |
|-----------------------------------|--------------------|---------|--------------------|---------|--------------------|---------|--------------------|---------|
|                                   | Univariate         |         | Multivariate       |         | Univariate         |         | Multivariate       |         |
|                                   | OR (95% CI)        | P value |
| Age per 10 years                  | 1.44 (1.03, 1.99)  | 0.032   | 1.48 (0.89, 2.45)  | 0.127   | 1.26 (0.80, 1.97)  | 0.321   |                    |         |
| Female sex                        | 1.99 (0.92, 4.33)  | 0.081   | 1.86 (0.76, 4.54)  | 0.176   | 1.05 (0.35, 3.19)  | 0.928   |                    |         |
| BMI per 5 kg/m <sup>2</sup>       | 0.59 (0.31, 1.12)  | 0.108   | 0.65 (0.31, 1.35)  | 0.247   | 0.58 (0.21, 1.63)  | 0.305   |                    |         |
| Hypogonadal stage                 | 0.99 (0.45, 2.19)  | 0.977   |                    |         | 2.82 (0.93, 8.51)  | 0.066   | 2.02 (0.60, 6.75)  | 0.256   |
| History of falls in the past year | 8.32 (0.97, 71.57) | 0.054   | 4.88 (0.48, 49.53) | 0.180   | 5.81 (1.16, 29.17) | 0.032   | 5.55 (0.86, 35.97) | 0.072   |
| Splenectomy                       | 1.68 (0.78, 3.59)  | 0.182   | 2.66 (0.97, 7.24)  | 0.056   | 1.30 (0.44, 3.88)  | 0.637   |                    |         |
| Iron chelating agents             |                    |         |                    |         |                    |         |                    |         |
| Deferiprone                       | 0.68 (0.32, 1.43)  | 0.307   |                    |         | 0.49 (0.16, 1.48)  | 0.205   |                    |         |
| Deferasirox                       | 2.61 (1.09, 6.23)  | 0.031   | 1.56 (0.58, 4.21)  | 0.378   | 2.98 (0.97, 9.14)  | 0.056   | 2.23 (0.66, 7.55)  | 0.197   |

Thavonlun et al. *Sci Reports* 2023

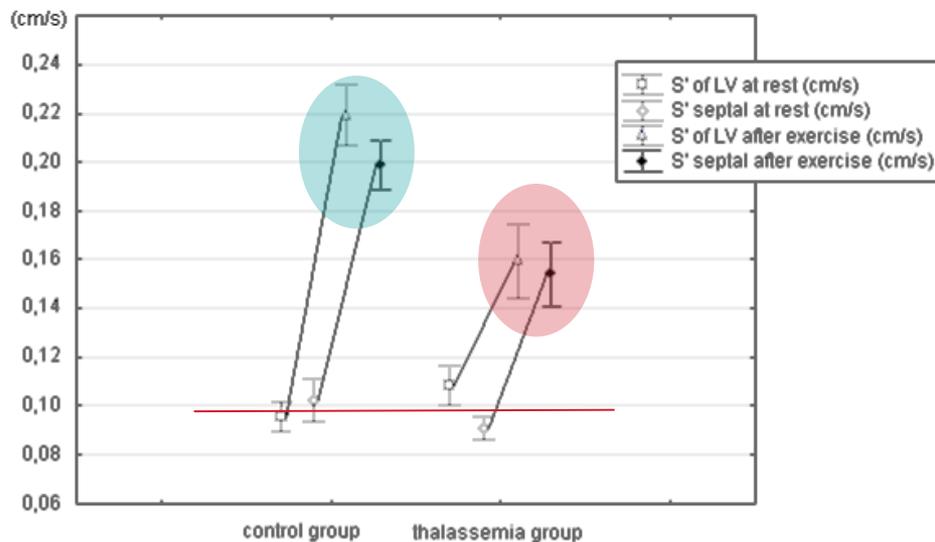
# 4- Anomalies démasquées à l'exercice physique

Cheung et al. *PLoS One* 2012



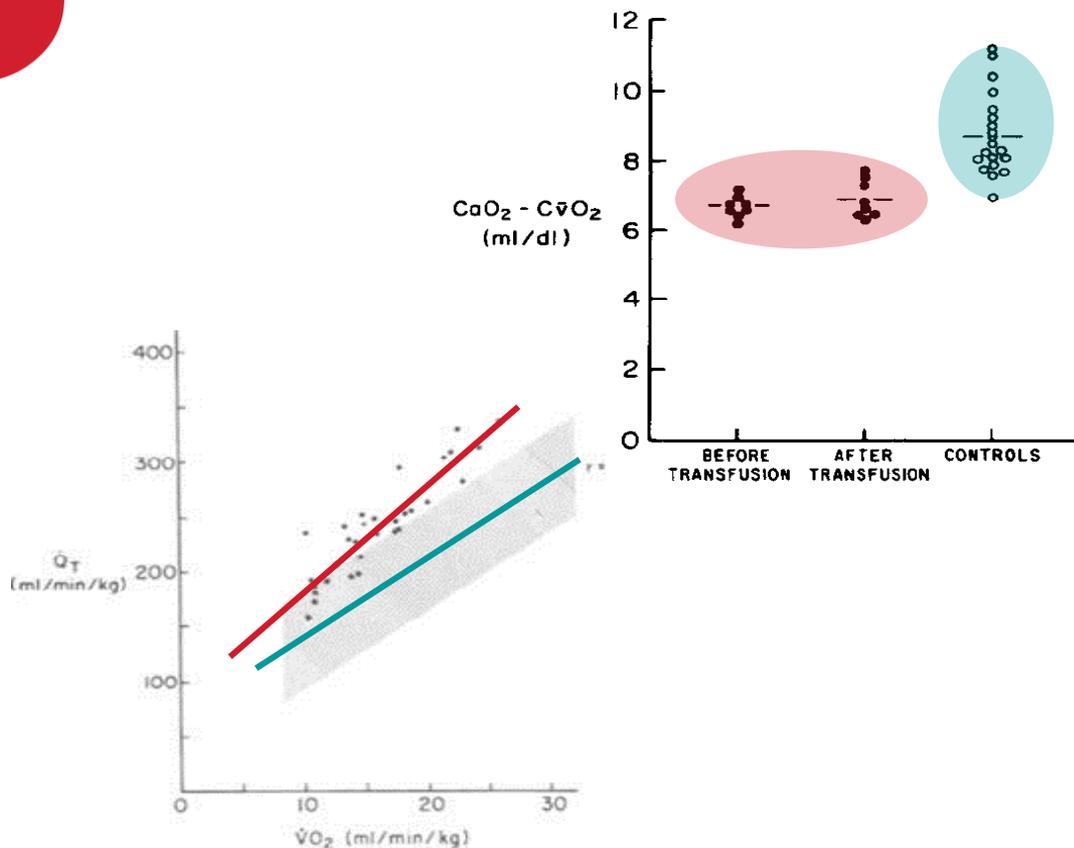
Scatter plots showing (a) baseline left ventricular systolic dyssynchrony index (SDI) at rest, (b) **SDI during exercise** when target heart rate was reached,

Barbero et al. *Mediterr J Hematol Infect Dis* 2012

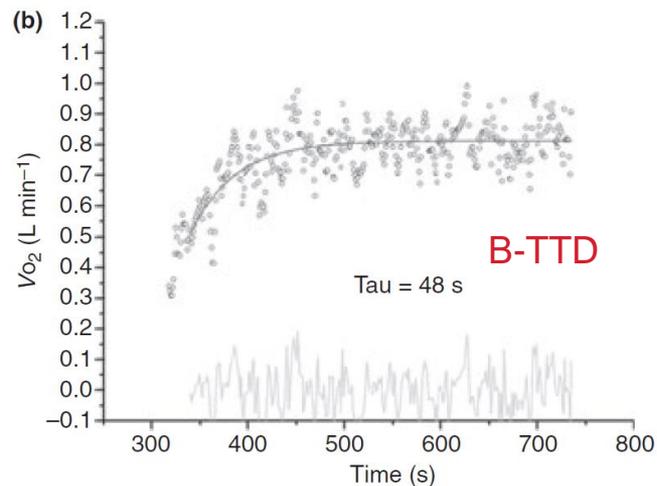
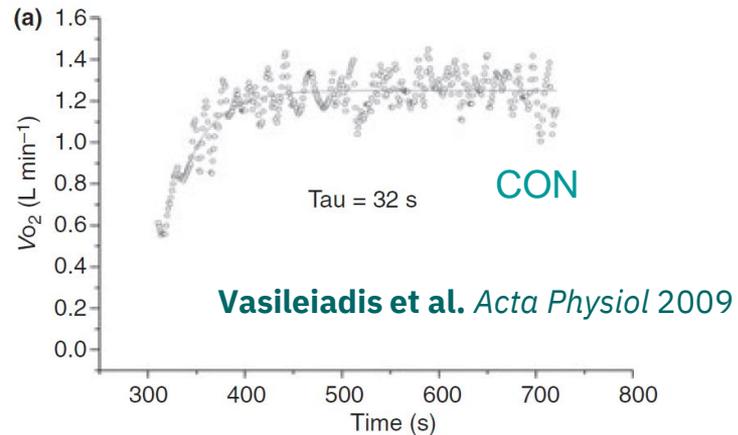


S' velocity of the Left Ventricle, at rest and after exercise, in patients with thalassemia (n=46) and in control group (n=39). **The difference of the velocity measured after the exercise in thalassemic patients versus control group was highly significant** ( $p < 0,0001$  for lateral wall's segment and septum).

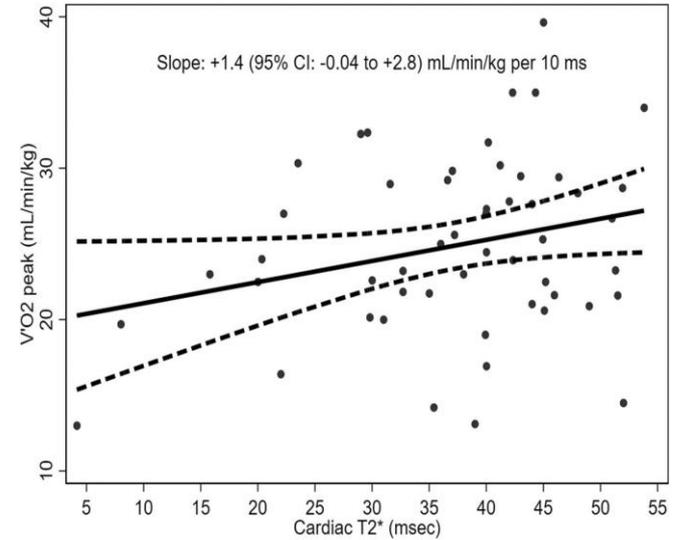
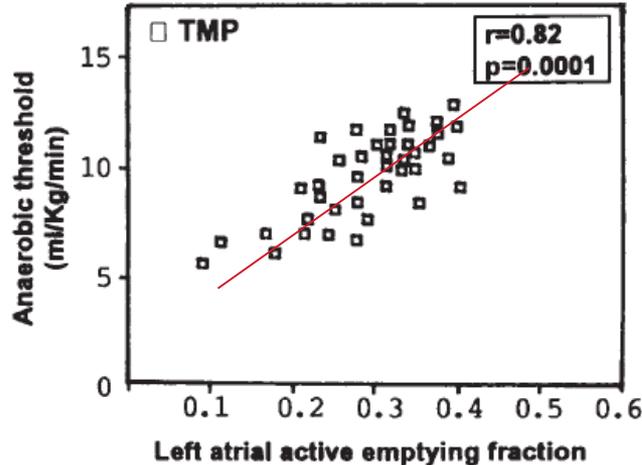
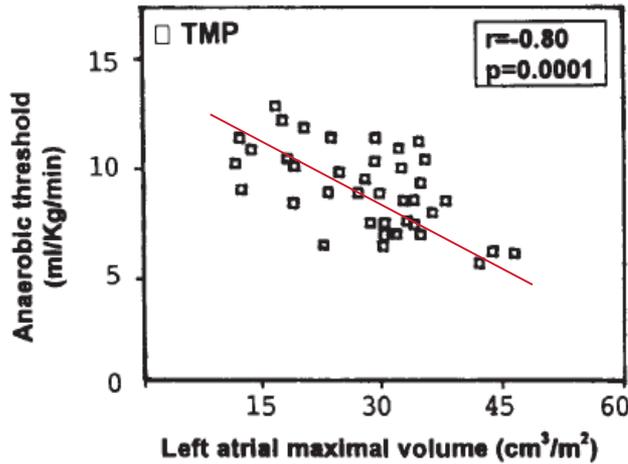
# 5- Réponses physiologiques “anormales” à l’effort



Grant et al. *Am Rev Respir Dis* 1987



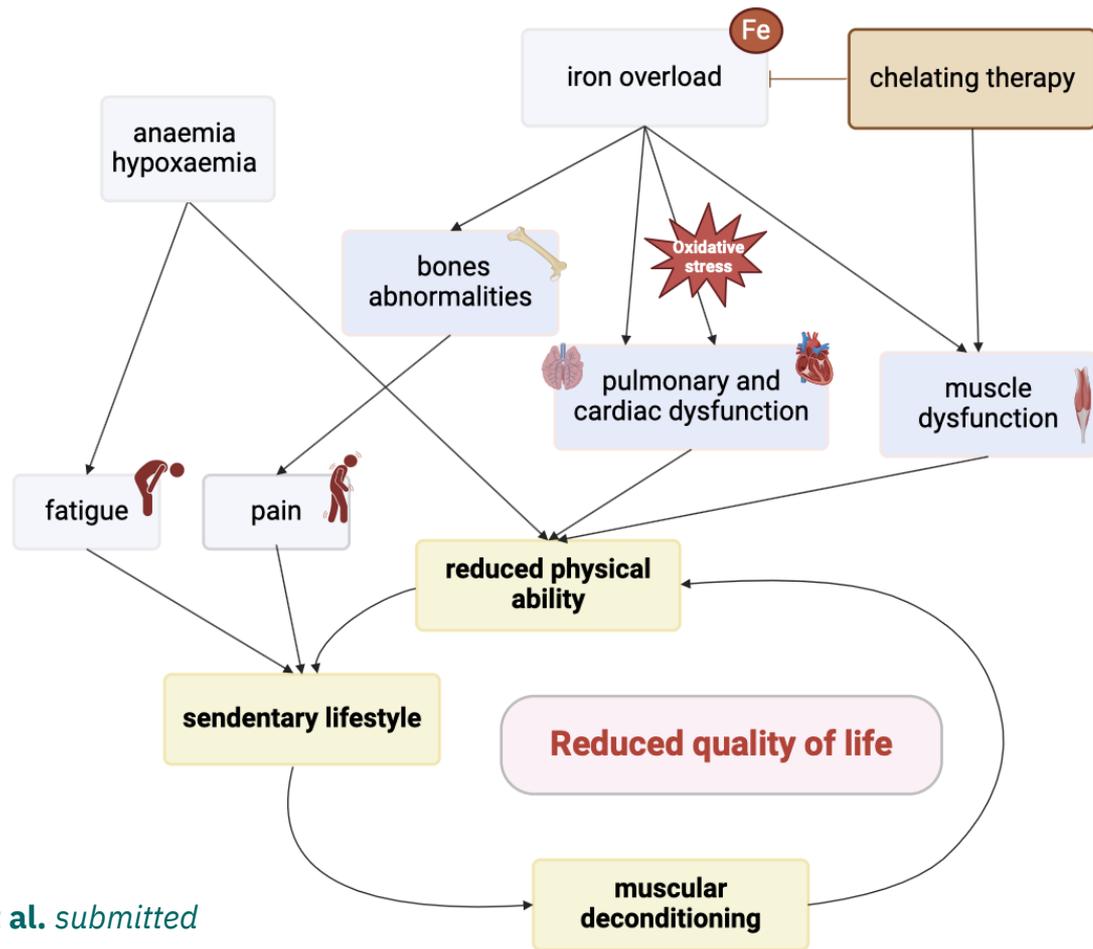
# 6- Limitations à l'effort



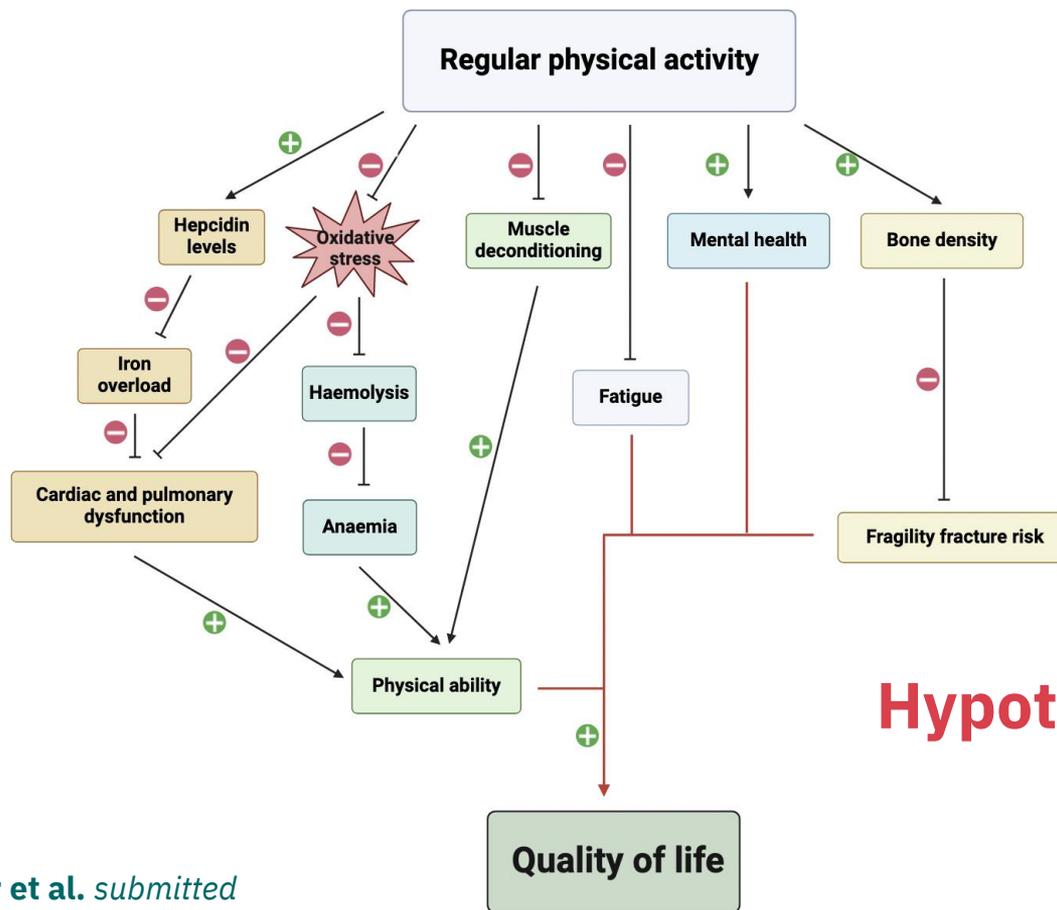
Piatti et al. *Ann Hematol* 2022

Trikas et al. *Am Heart J* 1998

# 7- Aptitude physique et qualité de vie

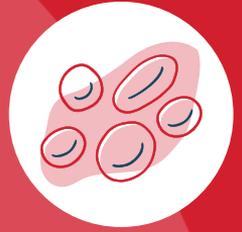


# 8- Rôles putativement bénéfiques de l'AP régulière



Hypothèses

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