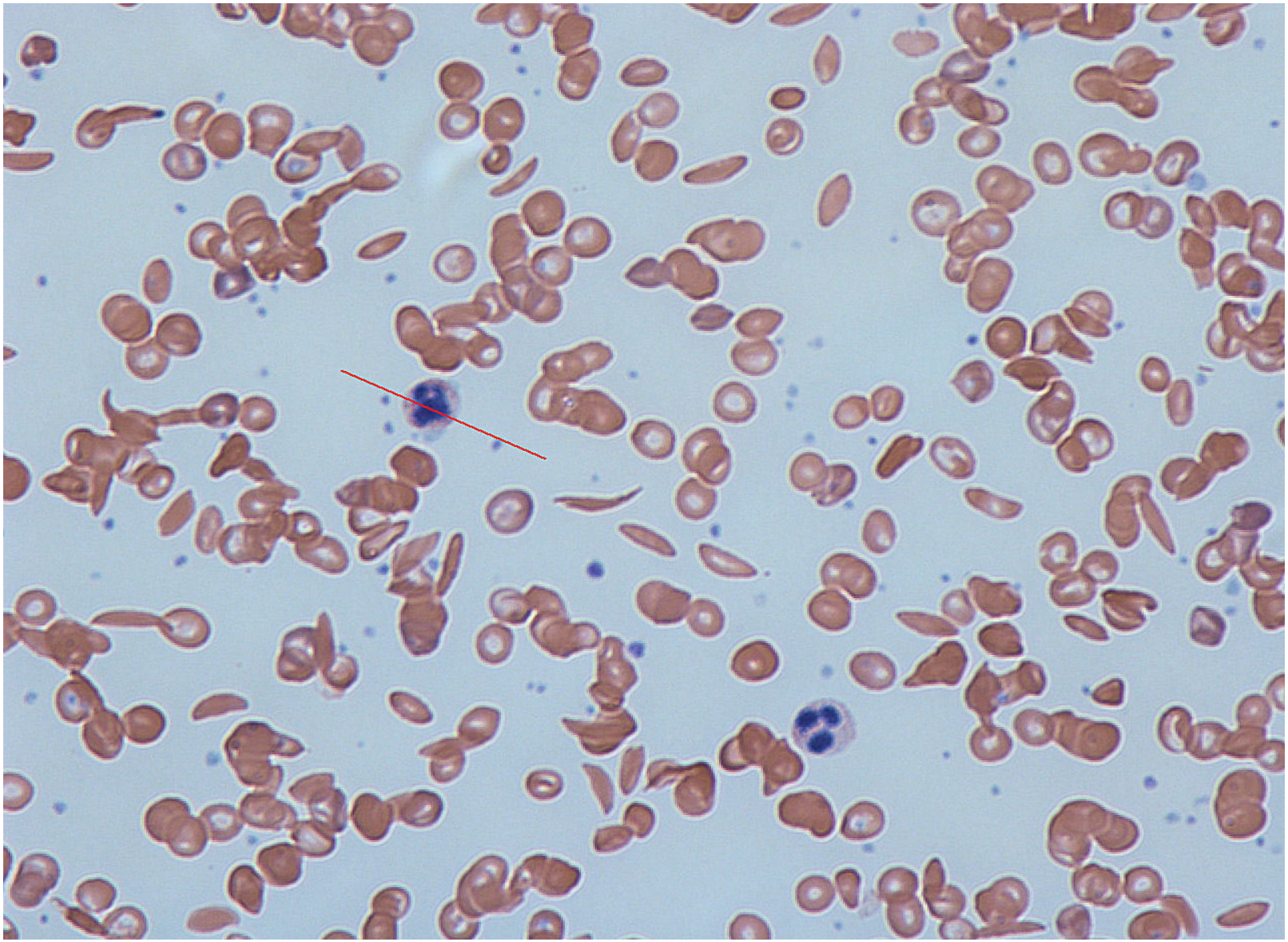
O coração na doença falciforme

Christiano Gonçalves Araujo

Sickle cell Disease

Introduction:

- Replacement of normal globin chains (Hb alfa 2, beta 2) for Hb SS (alfa 2, beta s 2, glutamic acid for valine), SC, SBeta, other rarer forms.
- Hb SS, unstable, low rates of O2, acidosis, hypothermia ... polymerization with formation of filamentous structures in erythrocytes: sickling
- Most common monogenic hereditary disease.



Pathophysiology

• Chronic anemia and hemolysis

• Increased viscosity (sickled erythrocytes, endothelium, leucocytes) with vaso-occlusive phenomena (ischemia / reperfusion).

Clinical manifestations

• Acute manifestations (+ children: VO, hemolytic, aplastic, kidnapping,) x chronic

•Chronic: systemic disease (spleen, MO, CNS, eyes, lungs, skin, kidneys …

Cardiovascular changes

- Dilation of the chambers , increased ventricular mass , presence of systolic murmurs

- LV dysfunction : Systolic - Diastolic

➢ pulmonary hypertension
➢ irregular heartbeat
➢ myocardial ischemia

Consequences :

1. Increased morbidity and mortality

2. Reduction FC

3. Marked worsening of prognosis

- Correlation with sudden death : 40 % of deaths as " sudden and unexpected ", much in the presence of any acute complications , cardiopulmonary causes x sepsis and addiction to narcotics , underdiagnosed target organ lesions.

Table 4 Independent correlates of increased tricuspid regurgitation velocity by multiple linear regression analysis

Variables	Odds ratio (95 % CI)	*p* value
Hb (g/dl)	1.1 (0.15–7.7)	0.916
Reticulocytic count (%)	1.98 (0.84–4.6)	0.117
Distance walked in 6-min walk test (m)	0.85 (0.74–0.98)	0.033[a]
Pretest O_2 saturation (%)	1.18(0.86–1.62)	0.291
SBP (mmHg)	1.22 (0.94–1.5)	0.123

CI confidence interval, *SBP* systolic blood pressure

The 6-min walk test: an independent correlate of elevated tricuspid regurgitant jet velocity in children and young adult sickle cell patients.

Hala Agha et al Ann Hematol DOI 10.1007/s00277-014-2030-3. Published online:22 february 2014.

Dilation of the chambers , increased ventricular mass, presence of systolic murmurs

- Progressive incidence - correlation with age, degree of anemia
- High prevalence
- Most common finding in cardiovascular examination.

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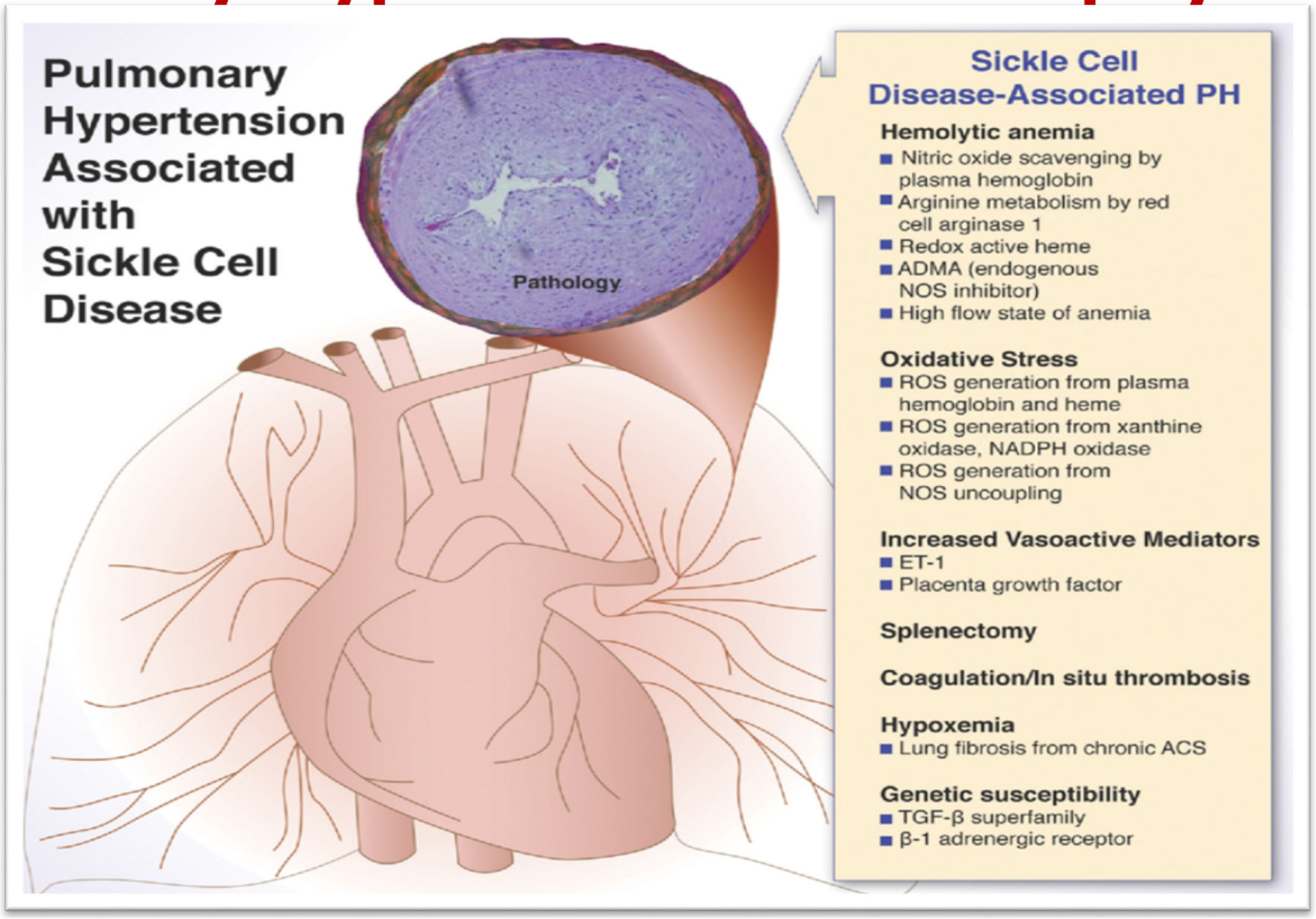
Updated Clinical Classification of Pulmonary Hypertension

4. Chronic thromboembolic pulmonary hypertension (CTEPH)
5. Pulmonary hypertension with unclear multifactorial mechanisms
   - 5.1 Hematologic disorders: chronic hemolytic anemia, myeloproliferative disorders, splenectomy
   - 5.2 Systemic disorders: sarcoidosis, pulmonary histiocytosis, lymphangioleiomyomatosis
   - 5.3 Metabolic disorders: glycogen storage disease, Gaucher disease, thyroid disorders
   - 5.4 Others: tumoral obstruction, fibrosing mediastinitis, chronic renal failure, segmental PH

Pulmonary hypertension : concept (mean PAP > = 25 mmHg) .
Pulmonary capillary pre x post (PCP < = 15 mmHg).

Diagnosis: Echocardiography - TRV , PASP . - Hemodynamic measurements of pressures . BNP – pro BNP. Prognostic importance.

Pulmonary Hypertension - Pathophysiology



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LV dysfunction : Systolic - Diastolic

- Prevalence - 18 % .
- Measurement of EF, other indices of systolic function .
- Diagnosing diastolic dysfunction , correlation with prognosis and PH.
- Association with iron deposition ?

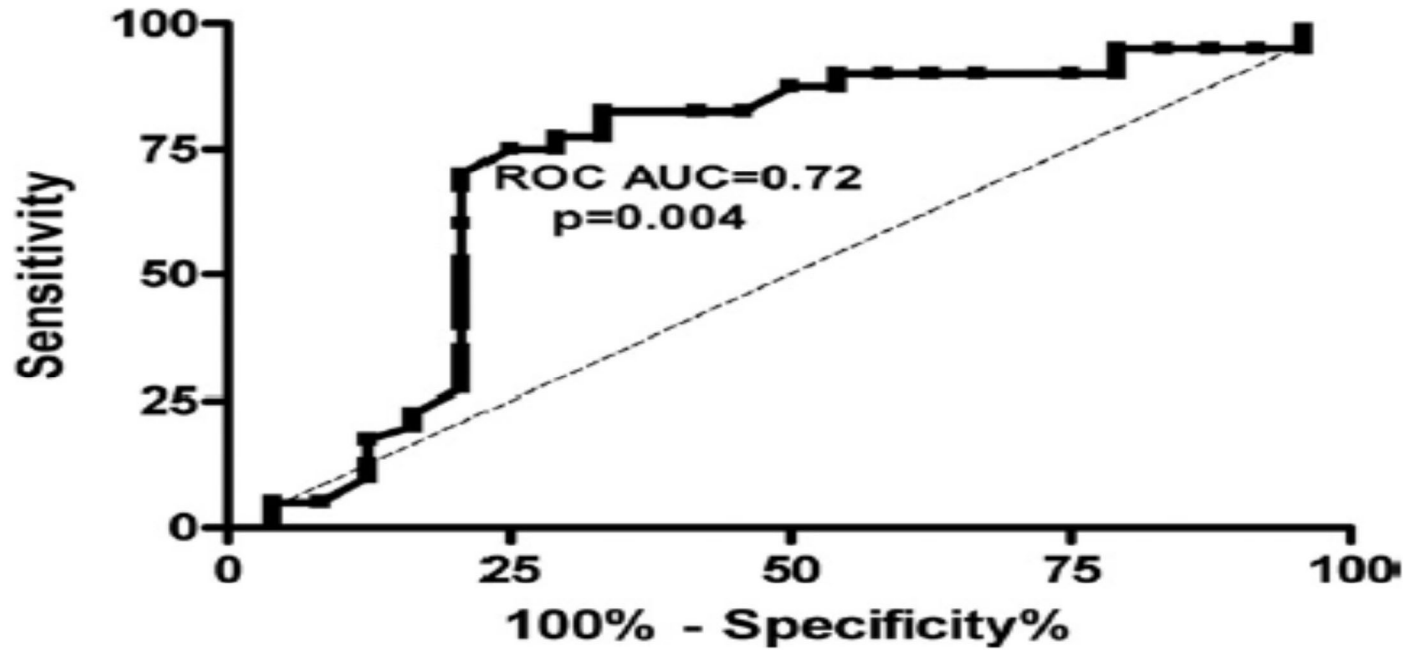

Figure 3 **Sensitivity and Specificity Analysis of Diastolic Dysfunction Measures in SCD Patients**

Receiver operating characteristic (ROC) curve for tissue Doppler lateral E/e' ratio to predict a pulmonary capillary wedge pressure >15 mm Hg by cardiac catheterization within 72 h in sickle cell disease patients. AUC = area under the curve.

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Irregular heartbeat

- Episodes detected by Holter or described during painful crises monitoring: prevalence of significant arrhythmias may reach 80% (half of them by ventricular origin)
- QT interval and ventricular ectopic
- Sudden death association?

Myocardial ischemia and coronary disease

- Myocardial ischemia associated with SCD**?**
- Necropsy findings and coronary angiography
- Underdiagnosed incidence?

Clinical manifestations

- Clinical diagnostic and propaedeutics:
- ECG
- Echocardiography
- Holter
- Chest X-ray
- Ergometry
- MRI
- Right CATE and angiography

Treatment: The underlying disease , other?

Follow-up

"The doctor may also learn more about the illness from the way the patient tells the story than from the story itself"

James B. Herrick.
Cardiologist, the first physician to describe the presence of sickle erythrocytes in the blood, its relationship with African descent and the presence of painful crises. 1910.