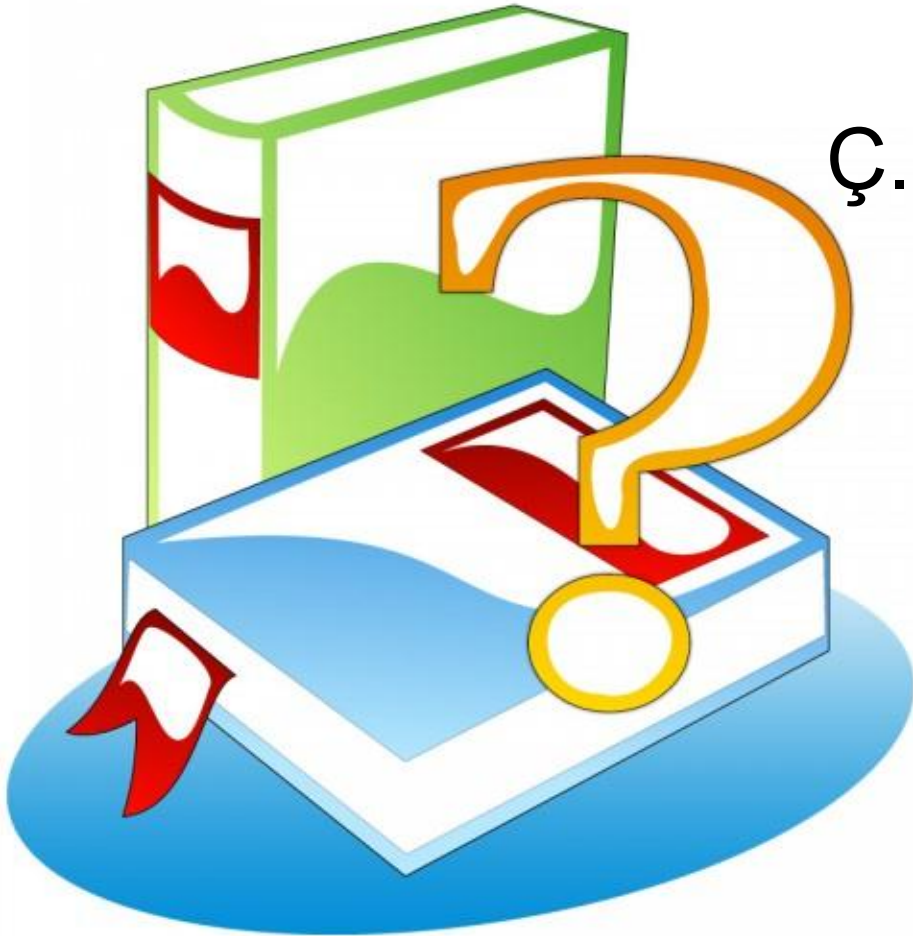


# ORAK HÜCRE ANEMİ KRİZ

Doç. Dr. Ayça Açıkalın  
Akpınar

Ç.Ü.T.F Acil Tıp AD



# Nedir bu orak hücre krizlerinden Çektiğimiz!!!!

- Hasta ağrılı krizde (ortalama 5-7 gün), geçmek bilmeyen ağrı
- Hastane personeli depresyonda, ağrı bir türlü giderilemediği için hasta yakını ajite
- Ağrılı kriz taklidi yapan meperidine bağımlısı hastalar kapıdan, bacadan meperidine temini için acilde doktorun peşinde

# Orak Hücre Anemi

## Amaç ve Sunu Planı

- Acilde Akut Vazooklüziv Ağrılı Krizlerin değerlendirilmesi ve yönetilmesi
- Ağrılı krizde ağrı tedavisi nasıl olmalı, güncel??
- Orak hücre aneminin özellikle ölümcül olabilecek diğer sistemik bulguları, tedavisi

# ORAK HÜCRE ANEMİ

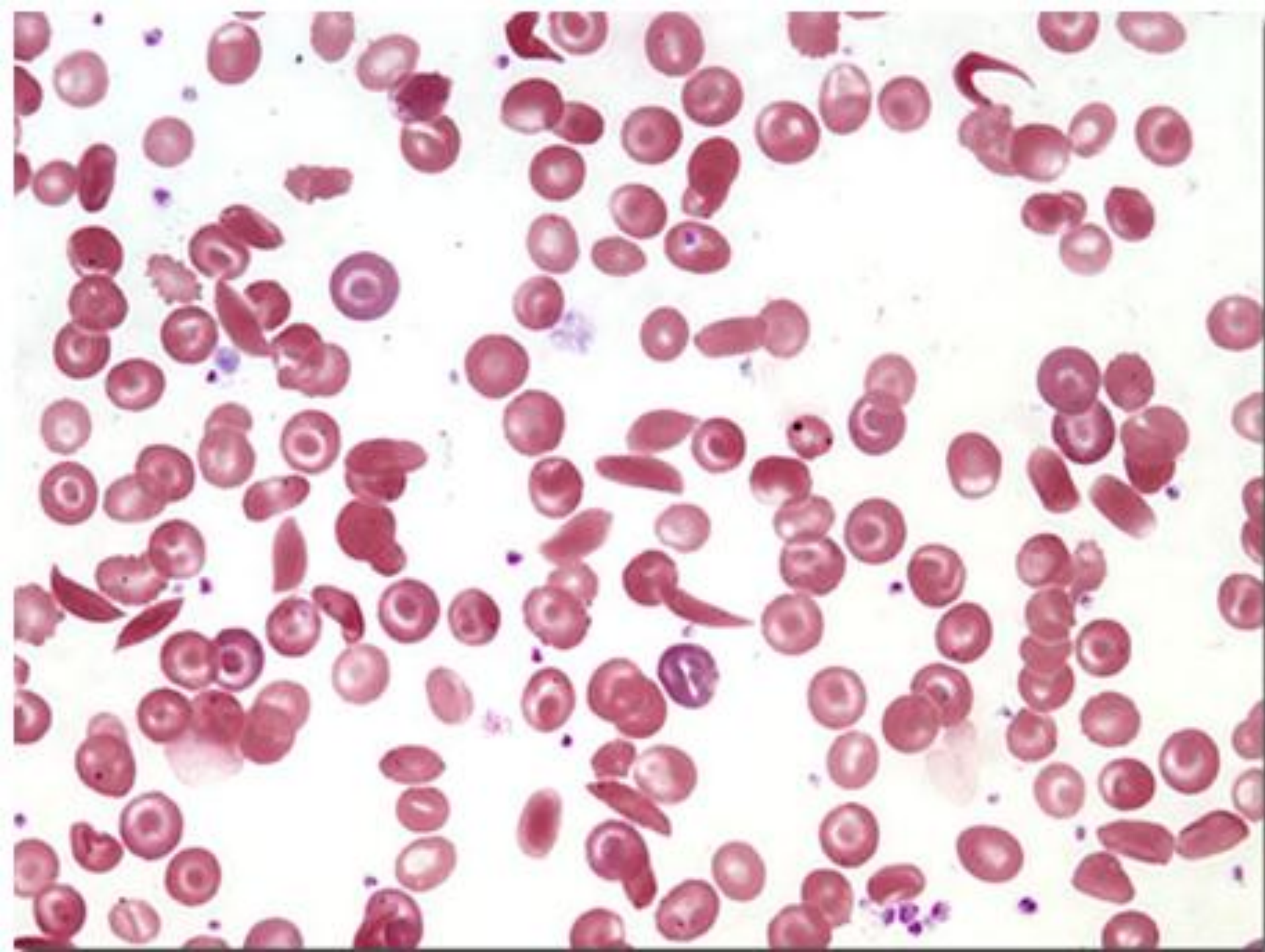
- **Dünyada en sık görülen hemoglobinopatilerden biri**
- **$\beta$ -globin zincirinin 6. pozisyonunda glutamik asitin valinle yer değiştirmesi sonucunda HBS oluşumu**
- **Otozomal resesif kalıtım**
- **Türkiye genelinde % 0.3-0.6 arasında**
- **Çukurova bölgesinde bazı yörelerde bu sıklık %3-44'e ulaşmakta**

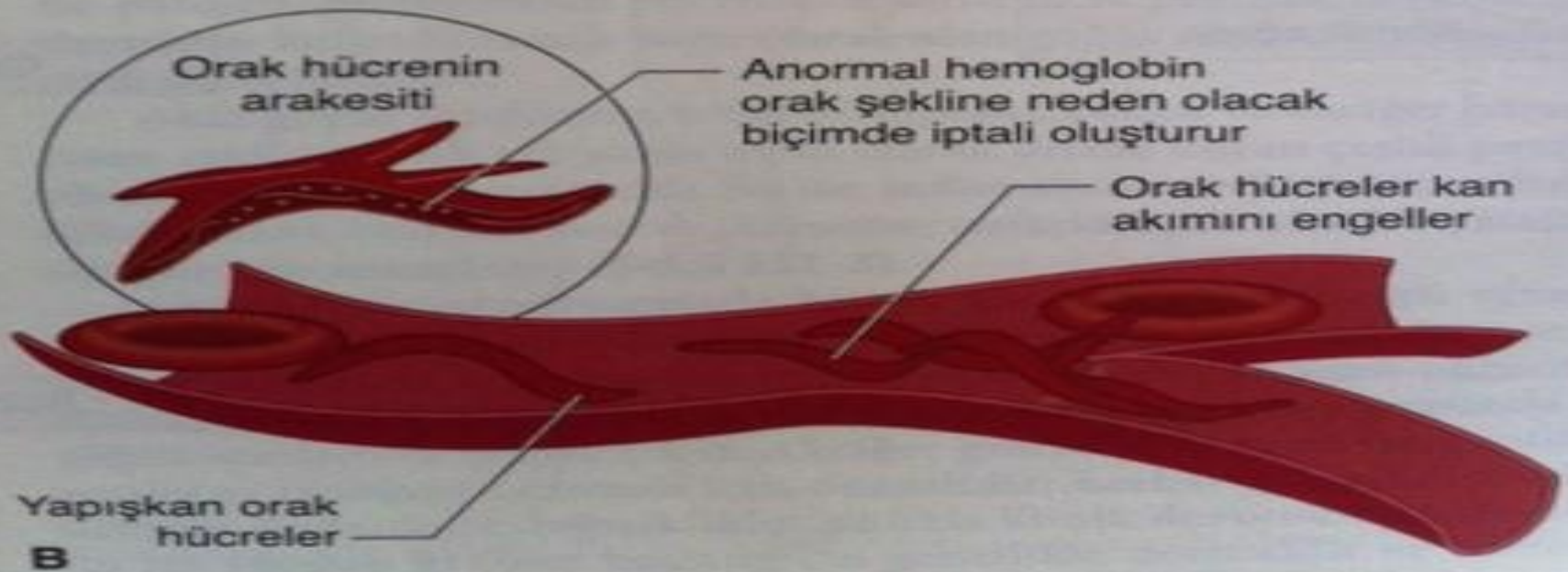
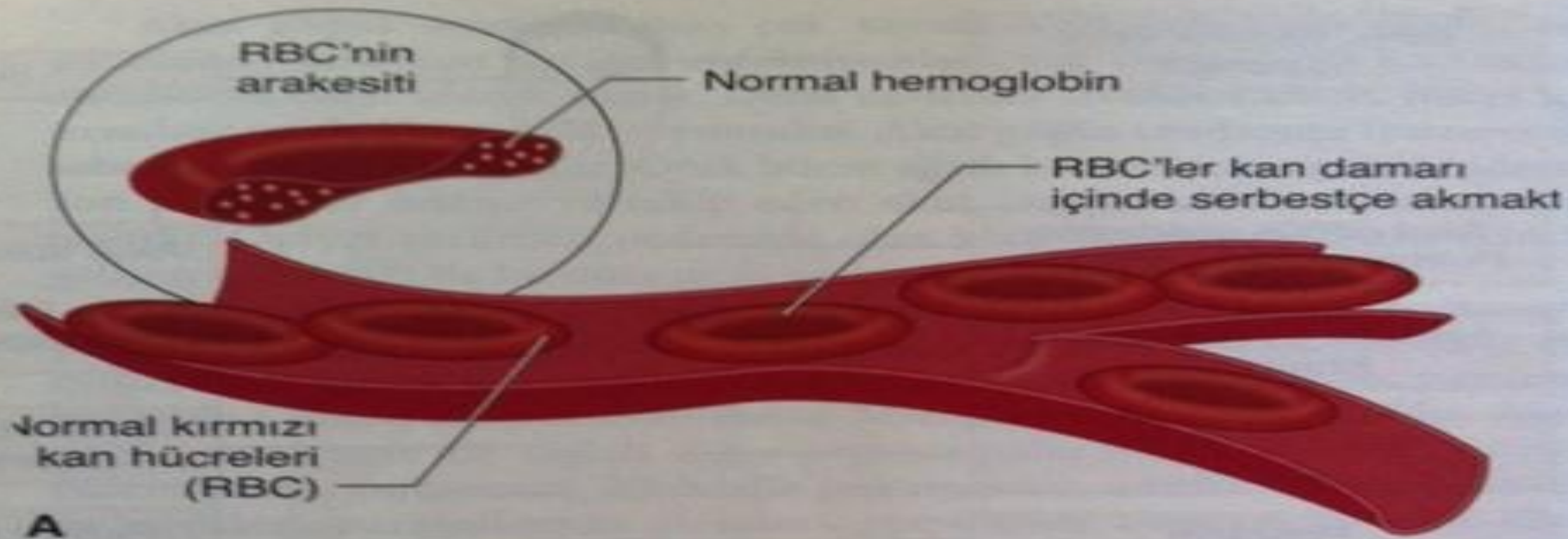
# Orak Hücre Anemi

- Dünya nüfusuna her yıl 6 milyon OHA taşıyıcısı, 1.2 milyon homozigot hasta katılmakta
- Orak Hücre bozuklukları tüm dünyada görülen hb bozukluklarının %70
- 1930'lı yıllarda yaşam beklentisi 14 iken günümüzde 50'li yaşlara çıkmıştır

# Orak Hücre Anemi

| Sendrom                             | Hemoglobin tipi  | Eritrosit içindeki yüzdesi | Tetramer içeriği (Globin zincirler)                                           |
|-------------------------------------|------------------|----------------------------|-------------------------------------------------------------------------------|
| Normal Erişkin                      | HbA              | 96-98                      | İki $\alpha$ -zincir ve iki $\beta$ -zincir                                   |
|                                     | HbA <sub>2</sub> | 3.0-3.5                    | İki $\alpha$ -zincir ve iki $\delta$ -zincir                                  |
|                                     | HbF              | 0.5-0.8                    | İki $\alpha$ -zincir ve iki $\gamma$ -zincir                                  |
| Orak Hücre Taşıyıcısı (Heterozigot) | HbA              | 60-85                      | İki $\alpha$ -zincir ve iki $\beta$ -zincir                                   |
|                                     | HbAS             | 35-40                      | İki $\alpha$ -zincir, bir normal $\beta$ -zincir, ve bir orak $\beta$ -zincir |
|                                     | HbF              | 0.5-0.8                    | İki $\alpha$ -zincir ve iki $\gamma$ -zincir                                  |
| Orak Hücre Anemisi (Homozigot)      | HbS              | 80-90                      | İki $\alpha$ -zincir ve iki $\beta$ -zincir                                   |
|                                     | HbA <sub>2</sub> | 2-4                        | İki $\alpha$ -zincir ve iki $\gamma$ -zincir                                  |
|                                     | HbF              | 2-20                       | İki $\alpha$ -zincir ve iki $\gamma$ -zincir                                  |





# Klinik

- Sonuç eritrositlerin ömrünün kısalması
- 120 günden ortalama 20 güne
- Kan viskozitesinin oraklaşma nedeniyle artması, mikrovasküler tıkanma
  - ( Sürekli hemoliz
  - ( Epizodik vasküler oklüzyon

İnme, iskemik veya  
hemorajik

Retinopati

Akut göğüs sendromu  
Pulmoner hipertansiyon

Akut splenik sekstrasyon  
krizi  
Dalak absesi  
Hipersplenizm

Hepatik sekestrasyon  
Kolelitiazis

Aku mezenterik  
iskemi

Orak hücre nefropatisi  
Böbrek yetmezliği

Priapizm

Femur başı avasküler  
nekrozu

Kemik ağrısı

Osteomiyelit

Bacak ülserleri



## 1. Damar tıkalıcı krizler

- Ağrılı krizler
- El-ayak sendromu
- Akut göğüs sendromu (AGS)
- Abdominal ağrılı krizler
- Akut Santral Sinir Sistemi Olayları
- Priapizm

## 2. Sekestrasyon Krizi

- Splenik Sekestrasyon Krizi
- Hepatik Sekestrasyon Krizi

## 3. Aplastik Kriz

## 4. Hiper Hemolitik Kriz

# Vazooklüziv Ağrı Krizi

- Acil Servise başvuruların en sık nedeni ağrılı krizler
- Oraklaşmış eritrositler çeşitli organlara giden kan akımını azaltarak iskemik ağrılara neden olur
- Hastaneye yatışın bir numaralı nedeni

# VAZO-OKLÜZİF AĞRI KRİZİ-PRESİPİTE EDEN NEDENLER

- Enfeksiyon
- Soğuk
- Dehidratasyon
- Rakım
- Gebelik
- Psikolojik Stres
- Fizyolojik stres
- Bilinmeyen nedenler

# Bu vakaların yönetimi nasıl olmalı?

- Agresif ağrı tedavisi !!!!!
- Hidrasyon
- Mevcut krizin nedeninin araştırılması
- Eşlik eden komplikasyonların varlığının araştırılması
- Etkin Tedavi

# Ağrılı Kriz Yönetimi-Hikaye

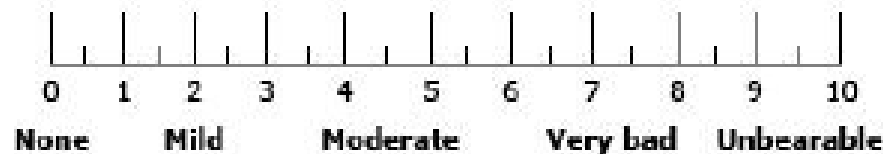
- Ağrının süresi, yeri
- Ağrı kesmek için kullandığı ilaçlar\*\*
- Ateş öyküsü
- Lokal şişlik kızarıklık öyküsü
- Akut atak tetikleyici faktör sorgulama\*\*

# Ağrılı Kriz Yönetimi- Fizik Muayene

- Ağrı düzeyinin derecelendirilmesi\*
- Ağrı bölgesinin ayrıntılı muayenesi
- Genel: Solunum sıkıntısı, solukluk, sarılık, döküntü
- Ayrıntılı solunum sistemi muayenesi
- Kalp muayenesi (KKY??)
- Karın Muayenesi (Organomegali, akut batın?, mezenter iskemi?)

### Monthly pain scale

**What does your pain feel like?**



Please mark in the circle of the number that most appropriately rates your pain level:

**0 = No pain**

**10 = Worst possible pain**

| 0 | 1 | 2 | 3 | 4 | 5 | 6 | 7 | 8 | 9 | 10 |
|---|---|---|---|---|---|---|---|---|---|----|
|---|---|---|---|---|---|---|---|---|---|----|

Right now

0 0 0 0 0 0 0 0 0 0 0

At its worst in the past month

[illegible]

At its least in the past month

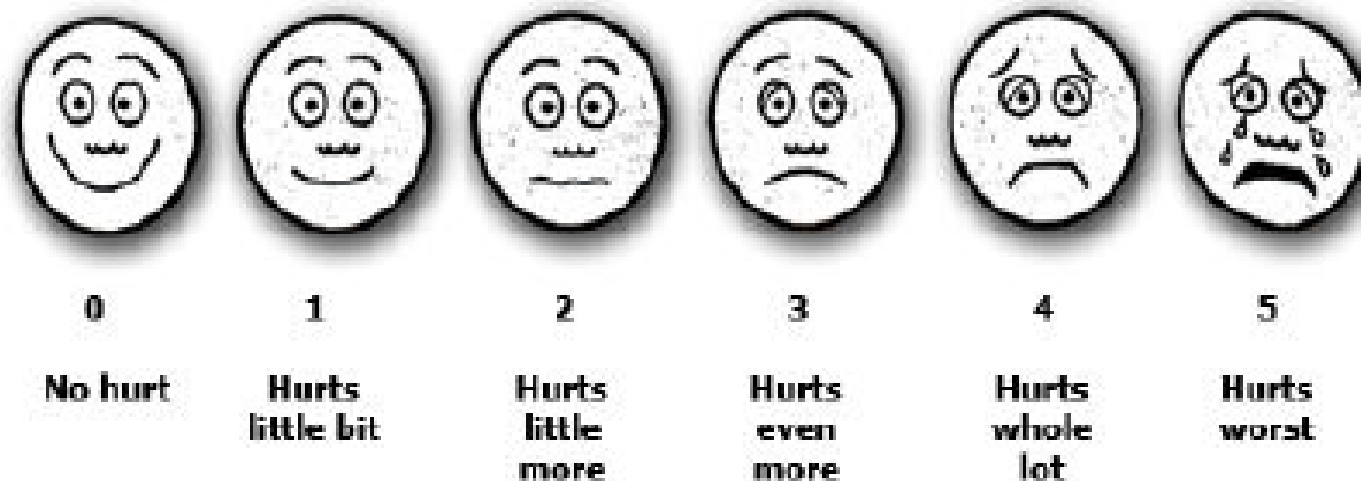
[illegible]

At its *average* in the past month

○ ○ ○ ○ ○ ○ ○ ○ ○ ○ ○ ○

## Wong-Baker FACES pain rating scale

---



Explain to the child that each face is for a person who feels happy because he has no pain (hurt) or sad because he has some or a lot of pain. Face 0 is very happy because he doesn't hurt at all. Face 1 hurts just a little bit. Face 2 hurts a little more. Face 3 hurts even more. Face 4 hurts a whole lot. Face 5 hurts as much as you can imagine, although you don't have to be crying to feel this bad. Ask the child to choose the face that best describes how he is feeling. Rating scale is recommended for persons age three years and older. Point to each face using the words to describe the pain intensity. Ask the child to choose the face that best describes own pain and record the appropriate number.

# Ağrılı kriz yönetimi-Yardımcı Tetkikler

- Tam kan Sayımı, Lökosit, Retikülosit,
- Klinik durumun gerektirdiği ek tetkiklerin planlanması

## SEQUENCE OF EVENTS IN THE SICKLE CELL PAINFUL CRISIS

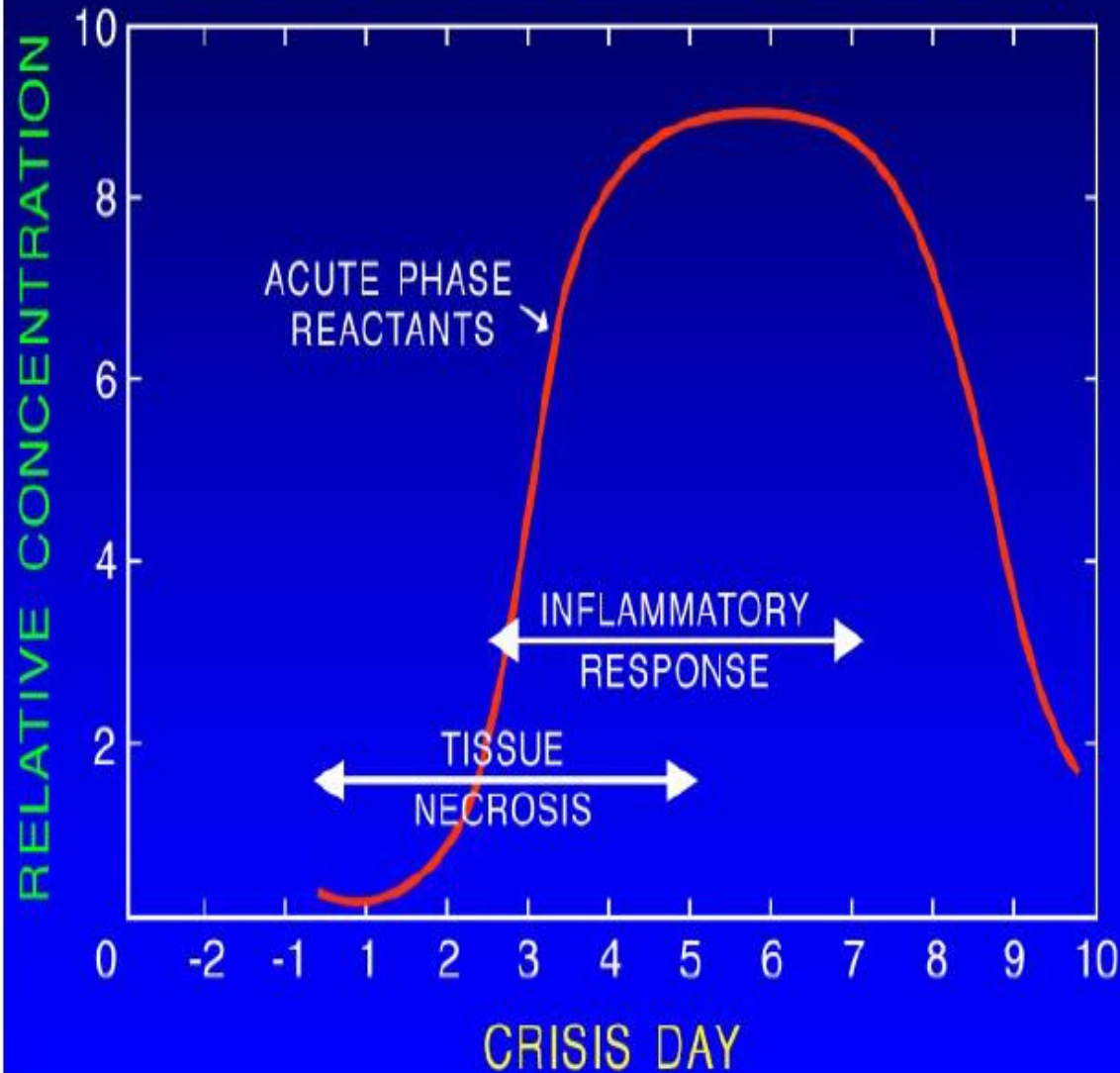
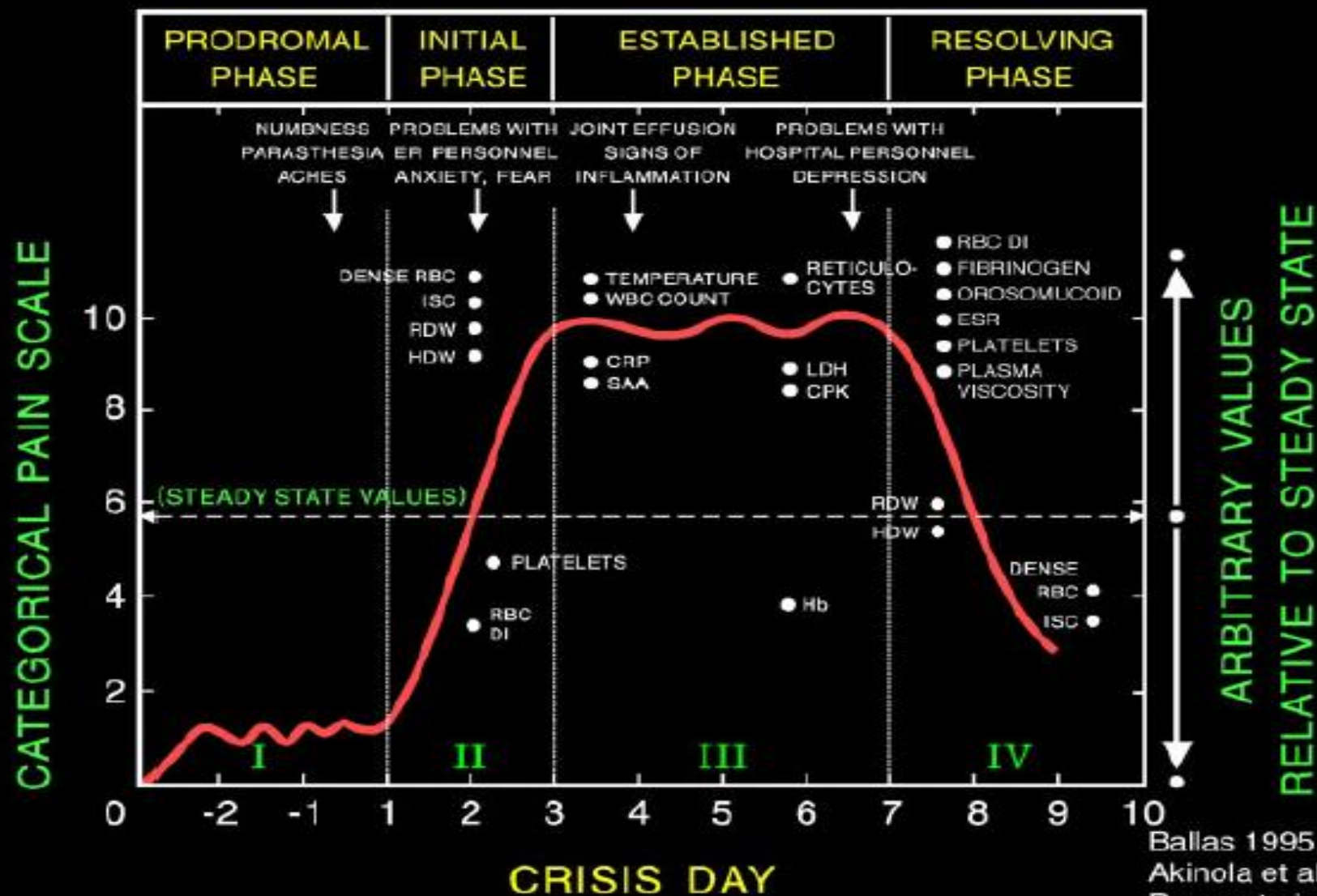


Figure 5. Sequence of events during the evolution of the prodromal phase of the painful crisis. Tissue necrosis consequent to ischemia elicits an inflammatory response that is associated with an increase in the serum level of acute phase reactants. Adapted from Ballas<sup>101</sup> with permission.

# CHANGES DURING THE PROGRESSION OF THE PAINFUL CRISIS



Ballas 1995, 1992  
 Akinola et al, 1992  
 Beyer et al. 1999  
 Jacob et al. 2005

**Table 1. Changes in objective signs during the evolution of the sickle cell painful crisis**

| Podromal phase    | Initial phase     | Established phase | Resolving phase   |
|-------------------|-------------------|-------------------|-------------------|
| Decreasing        | Decreasing        | Peak              | Peak              |
| RBC deformability | RBC deformability | Temperature       | Fibrinogen        |
|                   | Platelets         | WBC count         | Orosomucoid       |
| Increasing        |                   | Dense cells       | ESR               |
| Dense RBC         | Increasing        | ISC               |                   |
|                   | Temperature       | RDW               | Decreasing        |
|                   | WBC count         | HDW               | Temperature       |
|                   | Dense cells       | Reticulocytes     | WBC count         |
|                   | ISC               | LDH               | Dense cells       |
|                   | RDW               | CRP               | ISC               |
|                   | HDW               | SAA               | RDW               |
|                   | ESR               |                   | HDW               |
|                   | LDH               | Nadir             | CRP               |
|                   | CRP               | RBC deformability |                   |
|                   | Fibrinogen        | Hb                | Increasing        |
|                   | Orosomucoid       |                   | RBC deformability |
|                   | SAA               | Increasing        | Plasma viscosity  |
|                   |                   | Fibrinogen        | Platelets         |
|                   |                   | Orosomucoid       |                   |
|                   |                   | Plasma viscosity  |                   |
|                   |                   | ESR               |                   |

# Ağrılı Kriz Yönetimi- Genel Yönetim

Oral analjezikler ile kontrol altına alınabilecek,  
yeterli sıvı alımı var

• evde tedavi

Ağrılar parenteral tedavi gerektiriyorsa ,  
Ağızdan sıvı alamıyorsa,  
Birlikte komplikasyon mevcutsa

• Hastaneye yatır

Tablo 16. Ateşi olan OHA'lı hastalarda hastaneye yatırılarak izleme endikasyonları

|                                                                                                                                                                                                                                                                          |
|--------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|
| 3 yaş altında ve ateşi 38.6 °C üzerinde olan tüm OHA'lı hastalar                                                                                                                                                                                                         |
| Ateşi 40 °C üzerinde olan tüm OHA'lı hastalar                                                                                                                                                                                                                            |
| Septik görünüm                                                                                                                                                                                                                                                           |
| Peteşi ve purpura varlığı                                                                                                                                                                                                                                                |
| Hipotansiyon                                                                                                                                                                                                                                                             |
| Santral venöz kateter varlığı                                                                                                                                                                                                                                            |
| Geçirilmiş streptokokus pnömoniye bağlı bakteriyemi öyküsü                                                                                                                                                                                                               |
| Aşağıdaki akut komplikasyonların varlığına bağlı belirtilerin olması; <ul style="list-style-type: none"><li>• Pulmoner komplikasyonlar</li><li>• Aplastik kriz</li><li>• Sekestrasyon krizi</li><li>• İnme yada diğer nörolojik bozukluklar</li><li>• Priapizm</li></ul> |
| Beyaz küre sayısının $30 \times 10^9/l$ üzerinde yada $5 \times 10^9/l$ altında olması                                                                                                                                                                                   |
| Trombosit sayısının $150 \times 10^9/l$ altında olması                                                                                                                                                                                                                   |
| Daha önceki Hb düzeyine göre düşüklük                                                                                                                                                                                                                                    |

# Sıvı Tedavisi

- 5-10 ml/kg 1 saatte yükleme
- Daha sonra günlük idamesinin 1-1,5 katı olacak  
23 saatlik sıvı idamesi
- Ortalama günlük 3000 ml sıvı (2000 ml/m<sup>2</sup>)
- Agresif hidrasyon AGS için risk faktörü

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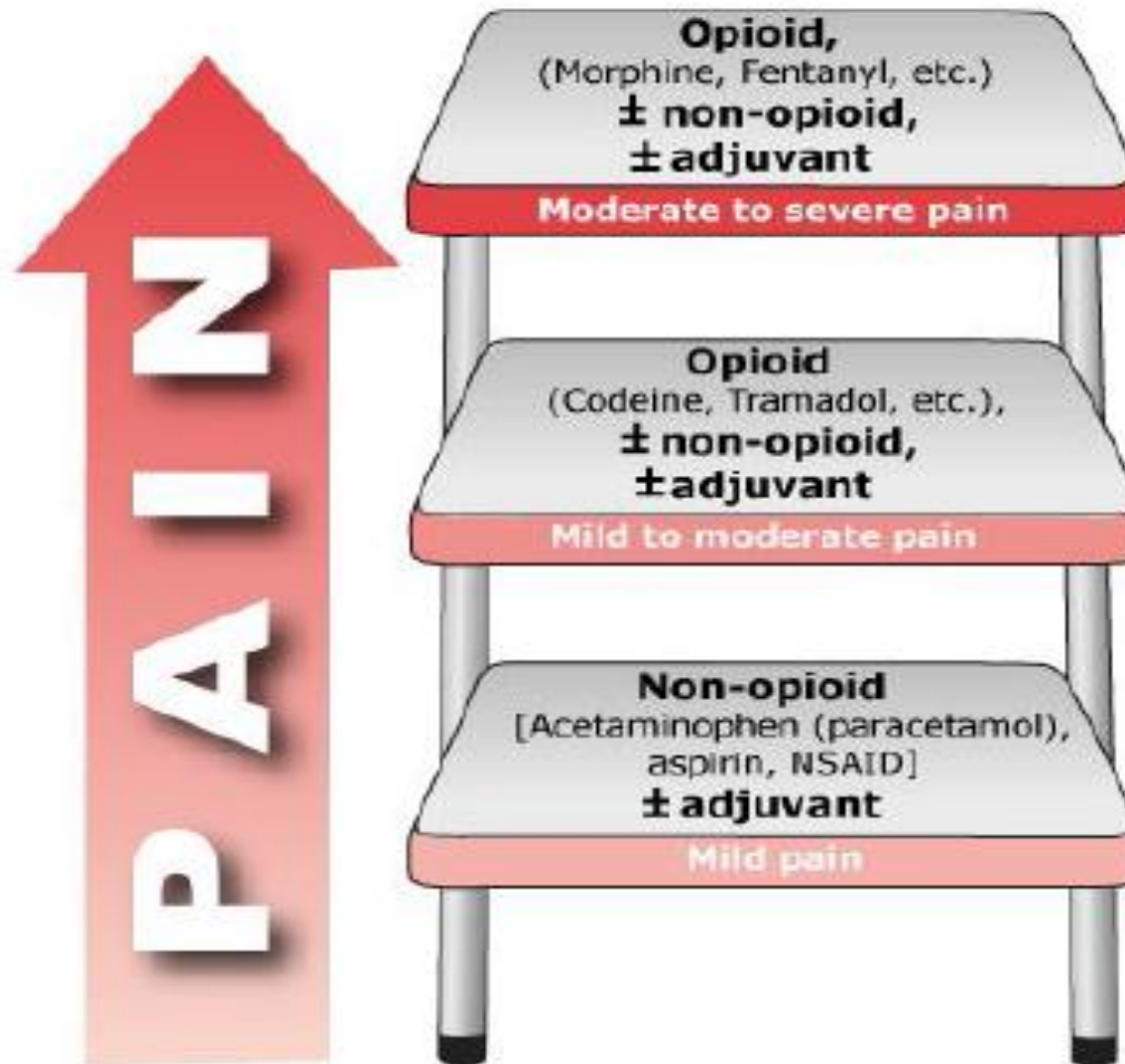
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**Deputy Editor**

Jennifer S Timauer, MD

## World Health Organization (WHO) analgesic ladder

---



# Narkotik Dışı Analjezikler

|               |                         |
|---------------|-------------------------|
| Aspirin (ASA) | 10mg/kg/4 saatte bir PO |
|---------------|-------------------------|

|              |                                                       |
|--------------|-------------------------------------------------------|
| Asetaminofen | 10-15mg/kg/4 saatte bir PO en yüksek doz: 40mg/kg/gün |
|--------------|-------------------------------------------------------|

|           |                                                        |
|-----------|--------------------------------------------------------|
| İbuprofen | 5-10mg/kg/6-8 saatte bir PO en yüksek doz: 40mg/kg/gün |
|-----------|--------------------------------------------------------|

|           |                                               |
|-----------|-----------------------------------------------|
| Naproksen | 5mg/kg/12 saatte bir PO en yüksek doz: 1g/gün |
|-----------|-----------------------------------------------|

|             |                            |
|-------------|----------------------------|
| İndometazin | 1-3 mg/kg/gün 3-4 dozda PO |
|-------------|----------------------------|

# Zayıf Narkotik Analjezikler

|        |                                               |
|--------|-----------------------------------------------|
| Kodein | 0,5-1mg/kg/4 saatte bir PO en yüksek doz 60mg |
|--------|-----------------------------------------------|

|           |                                 |
|-----------|---------------------------------|
| Oksikodon | 0,05-0,15mg/kg/ 4 saatte bir PO |
|-----------|---------------------------------|

|          |                                         |
|----------|-----------------------------------------|
| Tramadol | 50kg>50-100mg, 3 saat sonra tekrarlanır |
|----------|-----------------------------------------|

|           |                      |
|-----------|----------------------|
| Contramal | 1-2mg/kg İV infüzyon |
|-----------|----------------------|

# Kuvvetli Narkotik Analjezik

Morfin: IV bolus, IV inf., IM, SK, PO kullanımı mümkündür.

- 0,1-0,15mg/kg 3 saatte bir IV, en yüksek doz; 7,5-10mg

(birlikte parasetamol veya NSAİ başlanır. İki doz morfin ile ağrısı kontrol altına alınamayan hasta hastaneye yatırılır)

- IV devamlı infüzyon tedavisi; 0,04mg/kg/saat, 8 saatte bir 0,02mg/kg artırılarak en fazla 0,1mg/kg, idame tedavisi, 0,05mg/kg/3-4 saatte bir

- Hasta kontrollü analjezi; bazal infüzyon; 0,01-0,04mg/kg/saat, bolus doz: 0,01-0,04mg/kg,

# Dikkat

- Kreatinin klirensi 10-50 olan hastalara morfin hesaplanan dozun %75'i
- Kreatinin klirensi 10 ve altında olan hastalara morfin hesaplanan dozun %50'si verilir
- Renal yetersizliği olan hastalarda Fentanil iyi bir seçim
- Renal yetersizliği olan hastalarda NSAİ kontrendike

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# Morfin yan etkileri

- Bulantı-kusma : Ondansetron
- Kaşıntı: Hitrizin
- Konstüpasyon??
- Tüm yan etkiler için düşük doz naloksan kullanımı, ağrı kesici etkiyi azaltmadan yan etkileri gideriyor

ORIGINAL ARTICLE

# Treatment of the acute sickle cell vaso-occlusive crisis in the Emergency Department: a Brazilian method of switching from intravenous to oral morphine

Jessica Campos<sup>1</sup>, Clarisse Lobo<sup>1</sup>, Ana Maria Mach Queiroz<sup>1</sup>, Emilia Matos do Nascimento<sup>2,3</sup>, Carlos Bernardo Lima<sup>1</sup>, Gilberto Cardoso<sup>4</sup>, Samir K. Ballas<sup>5</sup>

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<sup>4</sup>Universidade Federal Fluminense – UFF, Niterói, Brazil; <sup>5</sup>Cardiac Foundation, Department of Medicine, Jefferson Medical College, Thomas Jefferson University, Philadelphia, PA, USA

## Abstract

**Objectives:** Describe the treatment of patients with vaso-occlusive crises (VOC) in a Brazilian emergency department (ED) and the successful switch from intravenous to oral morphine. **Patients and methods:** We analyzed records of 315 patients with sickle cell disease using two different protocols for pain: one in March 2010 prescribing intravenous morphine every 4 h throughout their stay, and another in March 2011 and 2012 prescribing one initial dose of intravenous morphine followed by equianalgesic doses of oral morphine every 4 h. Patients were triaged into three groups: mild, moderate, and severe VOC. The mild group was treated within 1 h after triage, the moderate within 30 min and the severe was treated immediately. Patients whose pain was not relieved within 6 h after the first dose of morphine were transferred into a different holding area of the ED where they continued to receive the same treatment for 48 h after which they were hospitalized if still in pain. **Results:** The number of patients who stayed <24 h in the ED increased significantly from 63 in 2010 to 87 in 2012, and the number of admissions decreased from 26 in 2010 to 10 in 2012. The incidence of acute chest syndrome decreased from 8.5% in 2010 to 1.9% in 2012. **Conclusion:** Patients treated with oral morphine stayed a shorter time in the ED, had more pain relief, were admitted less frequently, and had less acute chest syndrome. These differences may be due to environmental, cultural, psychological, and pharmacogenetic factors.

**Key words:** sickle cell disease; vaso-occlusive crisis; sickle cell pain; oral morphine; intravenous morphine

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# NIH Public Access

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## **Adult Emergency Department Patients with Sickle Cell Pain Crisis: Results from a Quality Improvement Learning Collaborative Model to Improve Analgesic Management**

Paula Tanabe, PhD, RN, MSN, MPH, John W. Hafner, MD, MPH, Zoran Martinovich, PhD, and Nicole Artz, MD

Schools of Nursing and Medicine, Duke University, (PT) Durham, NC; Department of Surgery – Emergency Medicine, University of Illinois College of Medicine at Peoria, and OSF Saint Francis Medical Center, (JWH) Peoria, IL; Division of Psychology, Mental Health Services and Policy Program, Feinberg School of Medicine, Northwestern University, (ZM) Chicago, IL; Loyola University Health System, (NA) Chicago, IL Previously: University of Chicago (NA), Chicago, IL

**Conclusions**—While the use of a learning collaborative and implementation of nurse-initiated analgesic protocols was not associated with improvement in time to administration of the initial analgesic, improvements in the decrease in the arrival to discharge pain score, and increased use of hydromorphone and the subcutaneous route, were noted in adults with SCD in the ED.

# Ketamine infusion for sickle cell pain crisis refractory to opioids: a case report and review of literature

Dipesh Uprety · Aurangzeb Baber · Maria Foy

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© Springer-Verlag Berlin Heidelberg 2013

**Abstract** This article reports a rare case of the use of low-dose ketamine infusion as an adjuvant to opioids to treat pain in sickle cell disease. A 31-year-old African-American male with history of sickle cell disease presented to the emergency department with complaints of chest tightness, multiple joint pain, and headache for 1 week. His vital signs and physical examination were unremarkable. His admission lab included hemoglobin of 8.4 g/dl, reticulocyte count of 16.3 %, bilirubin of 1.7 mg/dl, and LDH of 1,267 U/l. Chest X-ray showed middle and lower lobe opacity and interstitial thickening. He was treated for acute pain crisis and community-acquired pneumonia with intravenous fluids, supplemental oxygen, and intravenous levofloxacin. He was placed on fentanyl patient-controlled analgesia (PCA), oxycodone, ketorolac, and methadone with co-analgesic gabapentin and venlafaxine. Over the course of his hospitalization, his chest pain resolved, but the joint pains continued. He was then transferred to the ICU and was discharged a day later after 7 days of ketamine infusion. Ketamine is a noncompetitive antagonist at the *N*-methyl-D-aspartate (NMDA) receptor. This property has been

patient (5.9 %) developed serious side effect leading to discontinuation of the drug. A low-dose ketamine can be an option for pain control in sickle cell disease. Randomized trial is required to establish this benefit of ketamine over currently available therapies.

**Keywords** Sickle cell pain crisis · Ketamine · Opioid-induced hyperalgesia

Dear Editor,

Acute pain crisis is one the main causes of hospitalization in sickle cell disease. Opioids are the mainstay of therapy for pain related to sickle cell disease. However, some of the patients report continued pain because of tolerance and opioid-induced hyperalgesia. The postulated mechanism is the activation of *N*-methyl-D-aspartate (NMDA) receptor. This article reports a rare case of the use of low-dose ketamine infusion as an adjuvant to opioids to treat pain in sickle cell disease.

### *Clinical Note*

## The Role of a Low-Dose Ketamine-Midazolam Regimen in the Management of Severe Painful Crisis in Patients With Sickle Cell Disease

Qutaiba A. Tawfic, MBChB, FIBMS, Ali S. Faris, MBChB, FIBMS, and  
Rajini Kausalya, MD

*Department of Anesthesiology (Q.A.T., A.S.F.), The Ottawa Hospital, University of Ottawa, Ottawa, Ontario, Canada; and Department of Anesthesia (R.K.), Sultan Qaboos University Hospital, Muscat, Oman*

**Conclusion.** Low-dose ketamine-midazolam IV infusion might be effective in reducing pain and opioid requirements in patients with sickle cell disease with severe painful crisis. Further controlled studies are required to prove this effect. *J Pain Symptom Manage* 2014;47:334–340. © 2014 U.S. Cancer Pain Relief Committee. Published by Elsevier Inc. All rights reserved.

**Methods.** A retrospective analysis was performed with data from nine adult patients who were admitted to the intensive care unit with severe painful sickle cell crises not responding to high doses of IV morphine and other adjuvant analgesics. A ketamine-midazolam regimen was added to the ongoing opioids as an initial bolus of ketamine 0.25 mg/kg, followed by infusion of 0.2–0.25 mg/kg/h. A midazolam bolus of 1 mg followed by infusion of 0.5–1 mg/h was added to reduce ketamine emergence reactions. Reduction in morphine daily requirements and improvement in pain scores were the determinants of ketamine-midazolam effect. The *t*-tests were used for statistical analysis.

# Meperidine

rehospitalization rates for 408 children with SCD admitted with acute pain episodes found that having an outpatient visit was associated with lower rates of readmission to the hospital at two weeks and one month (hazard ratios [HR] 0.23, 95% CI 0.12-0.41 and 0.44, 95% CI 0.33-0.59, respectively) [70]. Similarly, in a single-center pediatric case control study, the greatest risk factor for readmission was no outpatient hematology follow-up within 30 days of discharge (OR 7.7, 95% CI 2.4-24.4) [69]. Among adults with SCD, the risk factors for readmission appear to be similar to those in children. This was illustrated in a single-center study in which approximately half of the patients admitted for acute painful episodes were readmitted within one month after discharge, and approximately 16 percent were readmitted within one week after discharge [71].

**Meperidine** — Meperidine is not recommended for the treatment of painful episodes of SCD, because multiple doses are associated with accumulation of normeperidine and central nervous system toxicity, including but not limited to mild twitching, multifocal clonus, and seizures.

# Bağımlılık

- Yurtdışında OHA'li hastaların madde bağımlılığı % 0-9
- Genel popülasyonda %6-9
- T.C'de bağımlılık oranı, özellikle meperidine bağımlılığı çok yüksek

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Acute pain management in adults with sickle cell disease

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# Gebede ağrılı kriz yönetimi

- Physical Exam
- Vital signs
- Pulse oximetry  
(Supplemental O<sub>2</sub> if SaO<sub>2</sub> is <95% on room air)

- Document fetal heart tones
- Continuous monitoring if >24 weeks

## Labs

- Complete Blood Count
- Comprehensive Metabolic Profile
- Reticulocyte count
- Urinalysis

Exclude/Treat Infection

- Admit to quiet location on bed rest
- Hydration: Oral or IV (0.5-1.0 liters of normal saline over the first hour then 125mL/hr)

## Assessment of patient's pain

- Allow patient input
  - Prior exposures
  - Specific side effects
  - Patient tolerance
- Pain assessments continued Q 30 min until pain is being controlled

\*Despite popular belief most patients are not drug seeking

## Aggressive Pain Relief (2 options)

1) Schedule opioid dosing (morphine/hydromorphone) Q 2-3 hrs with orders for breakthrough pain

2) Continuous opioid infusion (morphine/hydromorphone) with patient controlled anesthesia demand dosing every 20 minutes

## When to Wean

- No attempts until after 24 hours of adequate pain relief
- Wean by 10-20% daily with frequent pain assessments performed
- Convert to oral dosing when IV dosing is approximately equal to the patient's home regimen

## When to consider blood transfusion

-See Table 2

-Discharge home when pain is controlled on oral meds

Fig. 1 – Management of the pregnant patient who presents with painful sickle crisis.

# Hidroksiüre

| Hidroksiüre başlanması önerilen durumlar                                                                                                                                                                                                                  | Hastaya göre karar verilmesi gereken durumlar                                                                                                                            |
|-----------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|--------------------------------------------------------------------------------------------------------------------------------------------------------------------------|
| <ul style="list-style-type: none"><li>• Daktilit ve ağrılı krizlerin varlığı</li><li>• AGS varlığı</li><li>• Hemoglobin ve Hb F düzeyindeki düşüklük</li><li>• Beyaz küre ve LDH düzeyinde yükseklik</li><li>• Anormal Transkranyal USG varlığı</li></ul> | <ul style="list-style-type: none"><li>• Anormal beyin MRG (sessiz enfarkt)</li><li>• Nörokognitif fonksiyonlarda bozukluk</li><li>• Büyüme ve gelişme geriliği</li></ul> |

# Kan Değişimi (Exchange Transfüzyon)

- Tedaviye yanıt vermiyorsa kan transfüzyonu, KAN DEĞİŞİMİ düşünmek lazım.
- Ağrı tedavisi önemli, ağrılı krizde etkin ağrı tedavisi yapılması, akut göğüs sendromu ve mortaliteyi azaltmakta

# Akut Göğüs Sendromu

- Solunum sıkıntısı, ateş(>38,5), öksürük, eşlik eden göğüs, kosta, kemik, eklem ağrıları, hipoksi
- En sık 2-4 yaş
- Ağrılı vazooklüziv kriz sonrası 2. ölüm nedeni
- Yakın zamanda hastaneye yatış, entübasyon, AGS öyküsü
- Akciğer grafisinde yeni infiltrasyonlar

# Akut Göğüs Sendromu



# Tedavi

- Oksijen, Hidrasyon, Antibiotikler, Bronkodilatatörler
- Transfüzyon
- Tedaviye cevapsız, geçirilmiş AGS, entübasyon emboli şüphesi var ise kan değişimi düşünün!!!
- Kan değişimi, Hb oranı yüksek hastalarda daha etkin
- HbS oranını %30'un altına düşürülmeli
- Hidroksiürea (HbF miktarını artırır)
- İnhal NO (vazodilatasyon)

# Basit Transfüzyon

Semptomatik anemi

Akut nörolojik bulgu

Akut göğüs sendromu

Akut çoklu organ  
yetmezliği

Cerrahi öncesi

Akut splenik yada hepatik  
sekestrasyon

Sepsis ve menenjit

# Kan değişimi

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Akut nörolojik bulgu

Ağır akut göğüs sendromu

Akut çoklu organ yetersizliği

Cerrahi öncesi

Hipervolemi ve

hiperviskoziteden kaçınmak

Demir birikimini azaltmak

# Enfeksiyonlar

Orak Hücre  
anemide en sık  
mortaliteye ve  
morbiditeye neden  
olan sebep  
enfeksiyonlardır.



# Splenik Enfarkt

- Enkapsüle bakteriler tarafından ciddi enfeksiyon riski söz konusu
- Ateşi olan her hasta enfeksiyon açısından odak ve etken tespit edilene kadar geniş spektrumlu antibiotik almalı
- Pnömonoklara yönelik profilaksi almalı
- Grip aşısı, hem influenza aşısı

# Hemolitik Anemi

- OHA'de eritrositlerin şekilleri yüzünden kronik hemolitik bir süreç sözkonusu
- Bazal Hb: 6-9 gr
- Retikülosit sayısı: %5-15

# Aplastik Kriz

- Eritrosit üretiminde belirgin bir azalma
- Parvovirüs B-19, Folik Asit eksikliği
- Lökosit, trombosit normal
- Retikülosit(%0,5 altında)
- Kriz genelde 1 hafta içinde sonlanır
- Bu süreçte kan transfzyonu

# Nörolojik Tutulum

- İnme riski 200 kat artmıştır
- 20 yaşından önce OHA hastaların %10'u inme geçirir, %70-90 tekrar eder
- Geçirilmiş TIA
- HbF oranı %8'in altında olan hastalar
- AGS geçiren hastalar
- Sistolik kan basıncı yüksek hastalar
- Transkraniyal Dopplerde ön yada orta serebral arter kan akım hızının 200cm/sn veya yada daha yüksek olması
- Kan değişimi yapılmalı\*\*\*

