Centers for Disease Control and Prevention

Vital Signs

Preventing Sickle Cell Anemia Complications in Children

Screening and Treatment for Life-Threatening Problems Are Far Too Low

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20 Sickle cell anemia, which primarily affects Black people, can shorten life expectancy by more than 20 years.

50% Fewer than half of children 2–16 years received the recommended screening for stroke in 2019.

2 in 5 Only 2 in 5 children 2–9 years used recommended medication that can prevent sickle cell anemia complications in 2019.

Overview

Sickle cell anemia is the most severe form of sickle cell disease, a group of inherited red blood cell disorders causing unusually shaped, hard, and sticky red blood cells. These cells clump together, blocking blood flow carrying oxygen through the body. Sickle cell anemia, which primarily affects Black or African American people, is associated with a shorter life span and life-threatening complications that can affect all parts of the body. These complications cause pain and suffering. Sickle cell anemia is a common cause of childhood stroke.

There are screenings and treatments available to ease children's suffering from sickle cell anemia. Two recommended healthcare measures to prevent complications in children with sickle cell anemia are:

- Transcranial doppler (TCD) ultrasound screening, which identifies children with increased risk for stroke.
- Hydroxyurea therapy, which reduces the occurrence of several complications, including severe acute pain episodes and acute chest syndrome, which can result in lung injury and trouble breathing.

Far too few patients are receiving these potentially lifesaving prevention measures, recommended by an expert panel in 2014. Fewer than half of children aged 2–9 years (47%) and 10–16 years (38%) received a TCD ultrasound screening in 2019. Similarly, less than half of children aged 2–9 years (38%) and slightly over half of children 10–16 years (53%) used hydroxyurea. Managing sickle cell anemia in children is complex. These children face discrimination and multiple barriers to care. Racism and prejudice contribute to and worsen these barriers to care, making it harder to receive quality care. This leads to immense physical, emotional, and mental distress for children and their families.

Everyone can help improve care for people with sickle cell anemia by taking steps to address racism and prejudice. The healthcare system can promote tailored strategies to reduce barriers and increase TCD screening and hydroxyurea use among children with sickle cell anemia.

Read the full *MMWR*

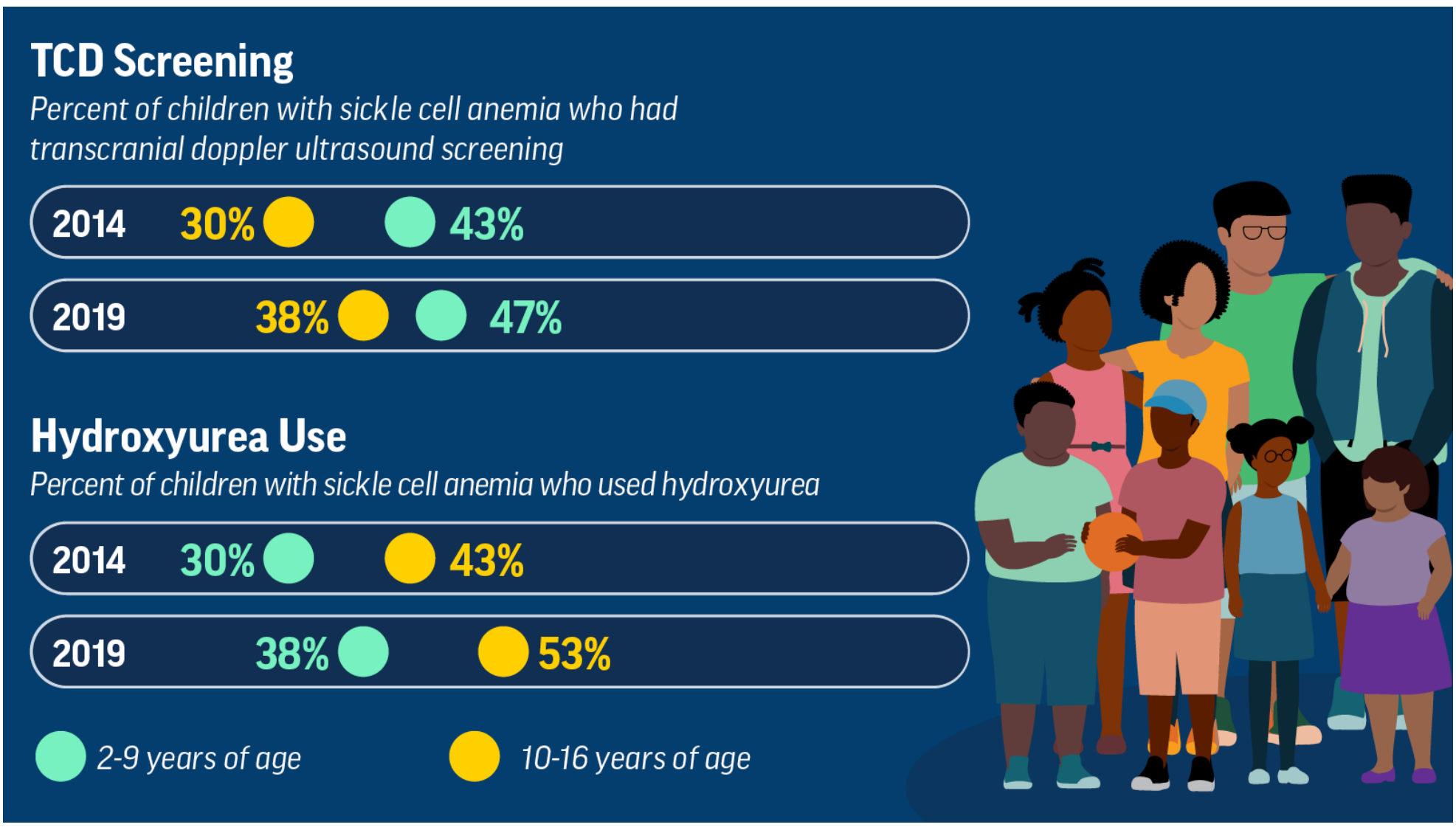
Challenges

Barriers to Care

- **Limited access to care:** There is a shortage of healthcare providers with expertise in treating patients with sickle cell anemia. Many providers may be unfamiliar with TCD screening and hydroxyurea guidelines.
- **Family and provider fears:** Fears of potential hydroxyurea side effects and uncertainty about whether the medicine will work can lead to low use.
- **Logistical issues with managing appointments:** Hydroxyurea therapy requires regular monitoring and laboratory visits. Availability of TCD screening appointments may be limited. Radiology centers that perform TCD screening may be far from where a child receives regular care.
- **Lack of care coordination:** Lack of timely information from radiology centers can make it hard for providers to track patients who are due for TCD screening.
- **Racism and prejudice:** Prejudice, discrimination, and bias against people with sickle cell anemia can lead to poor, unequal care. Policies leading to unequal opportunities in housing, employment, and health insurance access widen healthcare gaps and inequalities.

Too Few Children with Sickle Cell Anemia Are Getting Recommended Screening and Treatment

Despite recent improvements, many children with sickle cell anemia are still not receiving transcranial doppler ultrasound screenings and/or using hydroxyurea.



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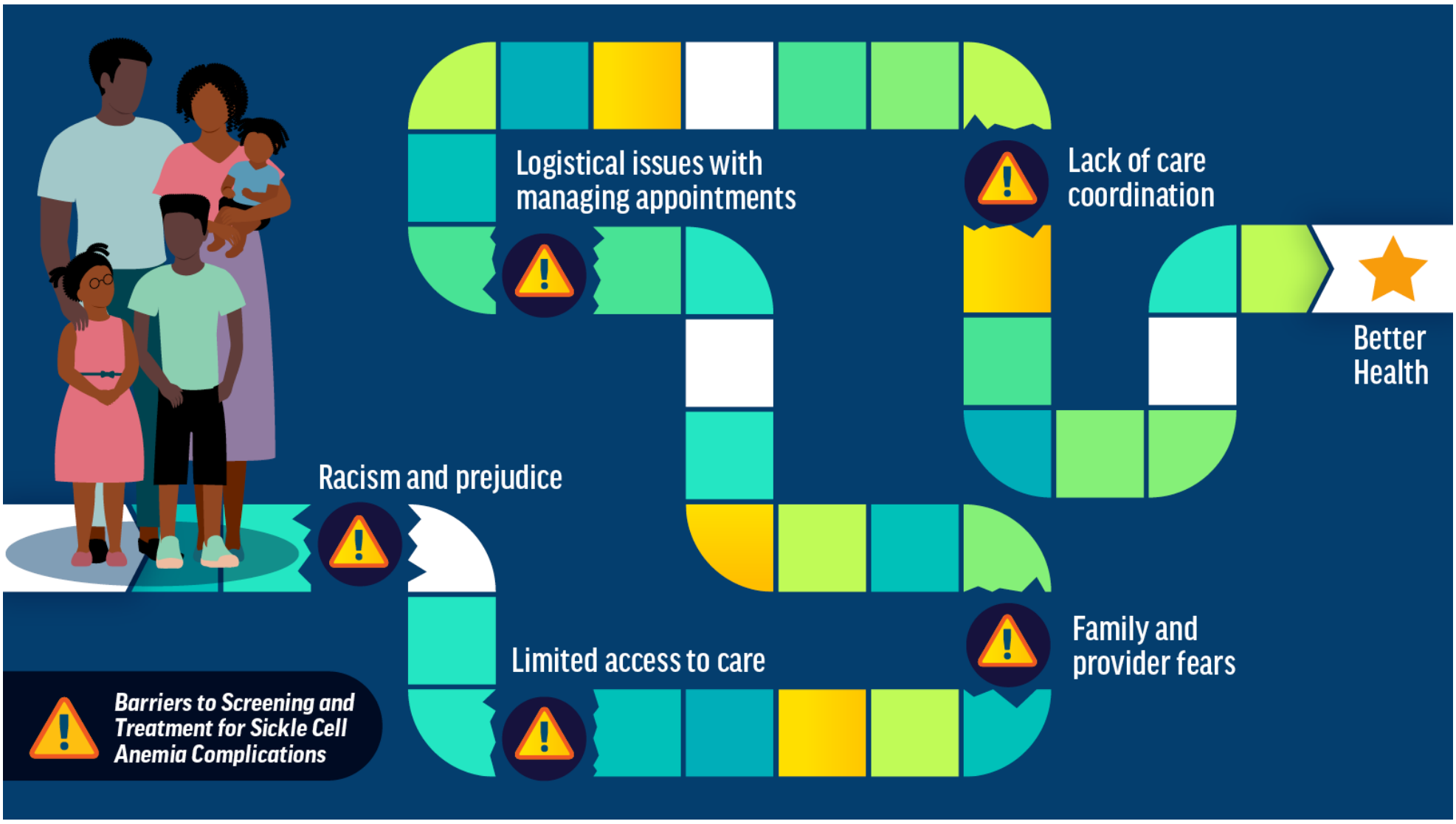
Children with Sickle Cell Anemia Face Barriers That Make It Hard to Get Care

While multiple paths can lead to better health, barriers to care widen health inequities for children with sickle cell anemia.



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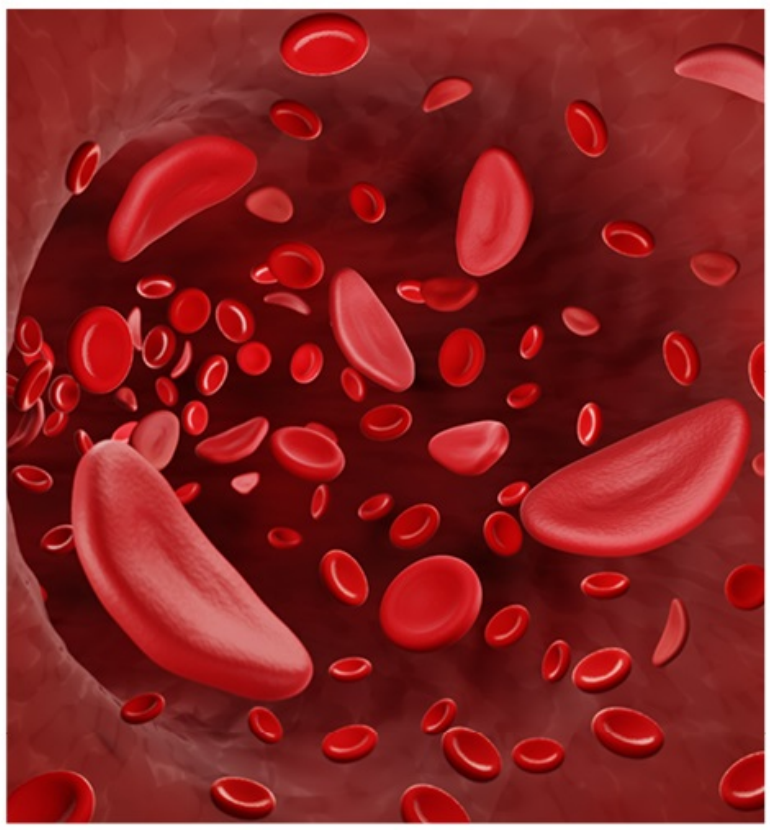
What Can Be Done

To Advance Health Equity

Preventing complications among children with sickle cell anemia includes recognizing where racism exists in the healthcare setting. It also means deciding to take steps to combat it. Healthcare providers can educate themselves, their colleagues, and the institutions they work with about the specialized needs of people with sickle cell anemia, including how racism is a barrier to better health. They can advocate for and listen to their patients to better understand their needs and support them on their journey to better health.

- **Work together to improve care:** Hematologists and other healthcare leaders can work with policy makers and advocates to reverse the impact of years of structural racism on sickle cell funding, research, and policy decisions.
- **Develop formal reporting systems:** Healthcare systems can develop formal reporting systems to document and respond to racist behavior and empower people with sickle cell anemia to safely report concerns about prejudice or inequity.
- **Increase education and training:** Healthcare education can emphasize training programs to increase knowledge of and capability in using the treatment guidelines most effective among people from racial and ethnic minority groups.
- **Use data to inform decision-making:** Local and state governments can use data to identify geographic gaps in care for people with sickle cell anemia and how to provide resources for those areas.



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Source: September 2022 *Vital Signs*

To Prevent Complications of Sickle Cell Anemia

Childhood stroke and other sickle cell anemia complications are preventable—not inevitable. Everyone has a role to play in prevention. Consider actions to increase TCD screening and hydroxyurea use among children with sickle cell anemia.

Increase TCD Screening

Healthcare Providers

- Implement actions that track TCD screenings and follow-up in electronic health records.
- Integrate screening into a single, comprehensive sickle cell visit whenever possible.
- Educate patients and families about screening.

Patients and Families

- Learn the importance of getting an annual TCD screening.
- Talk to your provider about TCD results and next steps if the results indicate a greater stroke risk.

Community-Based Organizations/Partners

- Develop patient and provider resources to improve understanding of the importance of annual TCD screening to prevent childhood stroke.
- Connect patients and families with resources and tools to schedule screening appointments, and support transportation needs by identifying financial assistance.

Increase Hydroxyurea Use

Healthcare Providers

- Become familiar with guidelines on prescribing hydroxyurea and studies showing the safety of hydroxyurea .
- Address practice barriers, such as lack of support staff or time to provide hydroxyurea counseling. Incorporate reminders into the electronic health record.

Patients and Families

- Learn about hydroxyurea and take as directed. Talk to your provider about the importance of regular monitoring and pros and cons compared with other treatments.

Community-Based Organizations/Partners

- Share patient resources about the benefits and safety of hydroxyurea [876 KB, 16 pages] .
- Develop tools to help patients take medication as directed (for example, mobile apps with reminders).
- Develop provider resources to increase familiarity with hydroxyurea safety and prescribing guidelines.

Footnotes and References

Related Pages and Resources

- *Vital Signs:* Media Statement – Many Children with Sickle Cell Anemia Not Receiving Lifesaving Screening and Treatment [English]
- *Vital Signs:* Media Statement – A muchos niños con anemia de células falciformes no se les hacen pruebas de detección y tratamiento que salvan vidas [Spanish]
- *Morbidity and Mortality Weekly Report (MMWR): Vital Signs:* Use of Recommended Health Care Measures to Prevent Selected Complications of Sickle Cell Anemia in Children and Adolescents — Selected U.S. States, 2019
- CDC: CDC Science Clips
- CDC: Sickle Cell Disease (SCD)
- CDC: Complications of Sickle Cell Disease
- CDC: Sickle Cell Data Collection Program
- CDC: Steps to Better Health for People with Sickle Cell Disease Toolkit
- CDC: Sickle Cell Disease Clinical Guidelines
- Agency for Healthcare Research and Quality: Transcranial Doppler Ultrasonography (TCD) Screening Among Children with Sickle Cell Anemia Toolkit
- National Institute for Children's Health Quality: Implicit Bias Resource Guide
- National Institute for Children's Health Quality: Sickle Cell Disease Treatment Demonstration Program
- University of Alabama at Birmingham: DISPLACE Trial
- *New England Journal of Medicine (NEJM)*: When Actions Speak Louder Than Words — Racism and Sickle Cell Disease
- National Academies of Sciences, Engineering, and Medicine: Addressing Sickle Cell Disease: A Strategic Plan and Blueprint for Action

- NHLBI, National Institutes of Health: Evidence-Based Management of Sickle Cell Disease: Expert Panel Report, 2014
- American Society of Hematology (ASH): SCD Resources for Clinicians
- ASH: Hydroxyurea for Sickle Cell Disease: Treatment Information from the American Society of Hematology [876 KB, 16 pages]
- Message from the ASH President on Diversity, Equity, and Inclusion *(June 9, 2020)*
- Message to the Membership from the Chair of the ASH Committee on Promoting Diversity *(July 12, 2021)*
- ASH Statement on Diversity, Equity, and Inclusion in Health Care *(February 9, 2022)*
- ASH Statement Addressing Diversity Equity and Inclusion in Hematology Research, Practice, and Training *(August 11, 2021)*
- ASH Clinical Practice Guidelines on Sickle Cell Disease

VITAL SIGNS RESOURCES

All CDC Vital Signs

Digital Media Tools

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