COMMONWEALTH of VIRGINIA

Department of Medical Assistance Services

CHERYL ROBERTS
DIRECTOR

SUITE 1300
600 EAST BROAD STREET
RICHMOND, VA 23219
804/786-7933
804/343-0634 (TDD)
www.dmas.virginia.gov

December 31, 2024

MEMORANDUM

TO: The Honorable Mark D. Sickles
Chair, House Health and Human Services Committee

The Honorable Ghazala F. Hashmi
Chair, Senate Education and Health Committee

The Honorable Rodney T. Willett
Chair, Joint Commission on Health Care

FROM: Cheryl Roberts
Director, Virginia Department of Medical Assistance Services

SUBJECT: Annual Review of Medications, Services, and Forms of Treatment for Sickle Cell Disease

This report is submitted in compliance with Section 32.1-331.06. of the Code of Virginia, which states:

C. By November 15 of each year, the Department of Medical Assistance Services shall prepare and submit a report that details the Department's findings from the annual review required by this section, as well as any recommendations to the General Assembly based upon those findings, to the Chairmen of the House Committee on Health and Human Services and the Senate Committee on Education and Health and to the Joint Commission on Health Care. The report shall be submitted for publication as a report document as provided in the procedures of the Division of Legislative Automated Systems for the processing of legislative documents and reports and shall be posted on the General Assembly's website and the Department's website.

Should you have any questions or need additional information, please feel free to contact me at 804-664-2660.

CR/wf
Enclosure

Pc: The Honorable Janet V. Kelly, Secretary of Health and Human Resources

Annual Review of Medications, Services and Forms of Treatment for Sickle Cell Disease Report

December 2024

Report Mandate:

Section 32.1-331.06. of the Code of Virginia states:

A. The Department of Medical Assistance Services shall conduct an annual review of all medications, services, and forms of treatment for sickle cell disease available to individuals with a diagnosis of sickle cell disease who are eligible for coverage under the state plan for medical assistance services. The purpose of the annual review is to determine (i) if the available covered medications, treatments, and services are adequate to meet the needs of enrollees with a diagnosis of sickle cell disease and (ii) whether the Department of Medical Assistance Services should seek to expand coverage for additional medications, treatments, or services.

B. When conducting the annual review required by this section, the Department of Medical Assistance Services shall solicit and consider input from the general public, with specific emphasis on input from persons or groups with knowledge and experience in the area of sickle cell disease treatment.

C. By November 15 of each year, the Department of Medical Assistance Services shall prepare and submit a report that details the Department's findings from the annual review required by this section, as well as any recommendations to the General Assembly based upon those findings, to the Chairmen of the House Committee on Health and Human Services and the Senate Committee on Education and Health and to the Joint Commission on Health Care. The report shall be submitted for publication as a report document as provided in the procedures of the Division of Legislative Automated Systems for the processing of legislative documents and reports and shall be posted on the General Assembly's website and the Department's website.

Annual Review of Medications, Services and Forms of Treatment for Sickle Cell Disease Report for 2024

In compliance with §32.1-331.06, DMAS conducted a review of medications, services and forms of treatment for Sickle Cell Disease (SCD).

For the Fiscal Year (FY) 2024 report, DMAS utilized claims and enrollment data, information provided by the MCOs, and input from providers, advocates, and Medicaid members.

Legislation

During the 2024 General Assembly session, several sickle cell-related bills were passed, including increasing sickle cell screening among adults (HB 255), creation of a voluntary sickle cell registry (HB 252), and requiring an annual review of treatments, covered services and medications by the State Medicaid Agency (HB 820).

§ 32.1-68 establishes a screening and treatment program within the Virginia Department of Health which includes education and post-screening counseling. The Code of Virginia instructs the Board of Health to adopt regulations to implement an adult and pediatric comprehensive sickle cell clinic network.

Sickle Cell Disease

Sickle Cell Disease (SCD) is a group of inherited red blood cell disorders. People with SCD have abnormal hemoglobin, called hemoglobin S or sickle hemoglobin, in their red blood cells. This abnormal hemoglobin causes the red blood cells to become rigid, sticky, and shaped like crescent moons or sickles. These sickle cells can block blood flow, leading to pain and other serious complications such as infections, acute chest syndrome, and stroke.

Individuals with sickle cell trait (SCT) have a mutation that causes them to make hemoglobin S instead of beta-globin they inherit one sickle cell gene and one normal gene. Although many individuals with SCT are asymptomatic, they can pass the trait on to their children. If both parents have SCT, there is a 50% chance that any child of theirs also will have SCT if the child inherits the sickle cell gene from one of the parents. If both parents have SCT, there is a 25% chance that any child of theirs will have SCD. There is the same 25% chance that the child will not have SCD or SCT. According to the American Society of Hematology, approximately one in 12 African Americans has SCT. Those of Hispanic, Middle Eastern, Asian, Indian and Mediterranean descent are also at risk for SCD and SCT.

Evidence-Based Treatment Guidelines

The **American Society of Hematology (ASH)** has developed comprehensive guidelines for the management of SCD, focusing on various aspects of the disease: [i]

1. **Pain Management**:
    - Use of nonsteroidal anti-inflammatory drugs (NSAIDs) and opioids for managing acute pain episodes.
    - Chronic pain management may include physical therapy, cognitive behavioral therapy, and medications.
2. **Hydroxyurea Therapy**:
    - Recommended for children and adults with frequent pain crises to reduce the frequency of these episodes and the need for blood transfusions.
    - Hydroxyurea can also help prevent acute chest syndrome and improve overall quality of life.
3. **Blood Transfusions**:
    - Regular blood transfusions are recommended for preventing stroke in children with abnormal transcranial Doppler ultrasound results.
    - Transfusions are also used to manage severe anemia and other complications.
4. **Stem Cell Transplantation**:
    - The only potential cure for SCD is a hematopoietic stem cell transplant (HSCT), typically from a matched sibling donor.
    - This option is considered for patients with severe

complications who have a suitable donor.

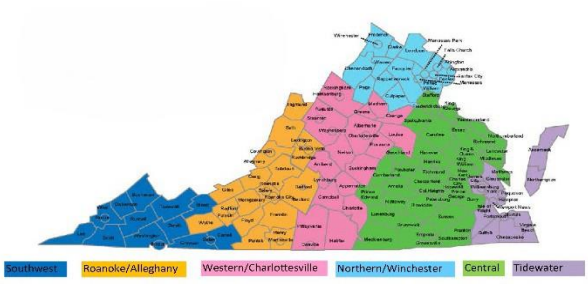
5. **Infection Prevention**:
    - Prophylactic penicillin is recommended for children until at least five years of age to prevent pneumococcal infections.
    - Vaccinations, including pneumococcal, meningococcal, and annual influenza vaccines, are crucial.
6. **Stroke Prevention**:
    - Regular screening with transcranial Doppler ultrasound in children to identify those at high risk for stroke.
    - Hydroxyurea and blood transfusions are used for primary and secondary stroke prevention.
7. **Management of Acute Chest Syndrome**:
    - Prompt treatment with antibiotics, oxygen therapy, and blood transfusions.
    - Pain management and incentive spirometry to prevent recurrence.

These guidelines are designed to improve the quality of life and outcomes for individuals with SCD by providing evidence-based recommendations for the prevention and management of complications.

Demographics

During SFY2024, there were 2,392 Medicaid and CHIP enrollees identified with a SCD diagnosis. Four thousand, five hundred and thirty-three Medicaid and CHIP enrollees were identified with a SCT diagnosis.

- **57%** of the population is female, **43%** male.
- **87%** identified as African American, **7%** Caucasian, **2%** Hispanic or Other race, **1%** Asian or Native American, and **less than 1%** Native Hawaiian or Pacific Islander or Bi-/Multi-racial.
- **42%** are less than 21 years old**, 28%** aged 21-34, **14**% aged 35-44, **9%** aged 43-54**, 5%** aged 55-64 and **3%** aged 65 and over.
- **2%** have limited Medicaid benefits and **less than 1%** have full Medicare coverage.
- **40%** reside in Tidewater, **32%** in Central, **17%** in Northern/Winchester, **7%** Charlottesville/Western, **4%** in Roanoke/Alleghany, and **less than 1%** in Southwest.



Treatment[ii]

Management of sickle cell anemia is typically aimed at preventing pain episodes, relieving symptoms and avoiding complications. Treatments may include medicines and blood transfusions.

Medicines

- **Hydroxyurea (Droxia, Hydrea).** Reduces the frequency of pain crises and possibly reduces the need for blood transfusions and hospital stays.

- **L-glutamine oral powder (Endari).** Helps in reducing the frequency of pain crises.
- **Crizanlizumab (Adakveo).** Helps reduce the frequency of pain crises in adults and in children older than 16 years.
- **Voxelotor (Oxbryta).** Decreases the risk of anemia and improves blood flow throughout the body.
- **Pain-relieving medicines.** Helps relieve pain during sickle cell pain crises.

Preventing infections

Children with sickle cell anemia should receive penicillin from about 2 months old to 5 years old, or longer to help prevent infections, such as pneumonia, which can be life-threatening to children with SCD. Adults who have SCD may need to follow a life-long penicillin regiment if they've had pneumonia or have had a splenectomy.

Recommended vaccinations are important for preventing childhood disease but are even more important for children with SCD because their infections can be life-threatening. Vaccinations should include pneumonia, meningitis, hepatitis B and an annual flu shot. Staying current on vaccinations is also important for adults with SCD.

Surgical and other procedures

- **Blood transfusions.** Blood transfusions are used to treat and prevent complications, such as stroke, in people with SCD. Transfusion increases the number of non-sickling blood cells and can help reduce the symptoms and complications of SCD.
- **Stem cell transplant.** Also referred to as a bone marrow transplant, the procedure involves replacing bone marrow affected by SCD with bone marrow from a matched donor, such as a sibling, who doesn't have SCD. Stem cell transplant is only recommended for individuals with significant symptoms and complications of SCD since the risks associated with the procedure are high and include death.
- **Stem cell gene addition therapy.** The individual's own stem cells are removed and a gene to produce typical hemoglobin is injected. The stem cells are then given back to the person in a process known as autologous transplant. This option may provide a cure for people with SCD without a well-matched donor.
- **Gene editing therapy.** Stem cells are removed from the body, and the sickling gene is changed, also called edited, to help restore the cells' ability to make healthy red blood cells. The treated stem cells are then returned to the body through the blood. This is called an infusion.

In December 2023, the U.S. Food and Drug Administration approved two treatments, Casgevy and Lyfgenia, the first cell-based gene therapies for the treatment of SCD in patients aged 12 years and older.

The Biden-Harris Administration announced action to increase access to SCD treatments through the Cell and Gene Therapy (CGT) Access Model developed by CMS. The CGT is designed to improve health outcomes, increase access to cell and gene therapies, and lower health care costs.

State-Certified CGT Centers

Name of Provider	State	Certified	Lyfgenia/Casgevey
Children's Hospital VCU	Virginia	✓	Lyfgenia
VCUHS Massey Comprehensive Cancer Center	Virginia	✓	Lyfgenia
Children's National Medica Center	Washington, DC	✓	Lyfgenia and Casgevy
Duke University Health	Durham, NC	✓	Lyfgenia and Casgevy
Tristar Centennial Medical Center	Nashville, TN	✓	Lyfgenia and Casgevy
Vanderbilt University Medical Center	Nashville, TN	✓	Lyfgenia and Casgevy

Virginia Medicaid Covered Services

97% of the Virginia Medicaid population diagnosed with SCD (2,331 members) has full coverage and access to all of the evidence-based treatment services recommended by the American Society of Hematology as well as the current treatments listed above. Virginia Medicaid enrollees diagnosed with SCD also have access to:

- medical benefits
- pharmacy benefits
- inpatient/outpatient hospital
- behavioral health services
- mental health and residential treatment services
- addiction and recovery treatment services
- long-term care services and supports (facility and/or community based)
- emergency transportation
- preventive services (such as vaccines and screenings)
- non-emergency transportation
- maternal and infant health services
- telehealth and telemedicine, and
- coverage in connection with clinical trials

Virginia Medicaid offers adult eye exams and treatment for the detection and prevention of SCD-related vision complications such as retinopathy. Virginia Medicaid managed care plans offer enhanced benefits which provide coverage for glasses, frames, or contacts.

Virginia Medicaid also offers routine dental care for children and adults through the Cardinal Care Smiles program offered through DentaQuest. Medicaid members living with SCD have access to dental services such as routine cleaning, exams, x-rays and fillings as well as gum related treatments, dentures, and root canals.

Virginia Medicaid covers audiology services that are provided as an inpatient, outpatient hospital service, outpatient rehabilitation agencies, or home health service to detect and

treat hearing loss associated with SCD.

Medicaid managed care organizations provide further support services for Medicaid enrollees diagnosed with SCD through:

- Care management services that utilize a whole health approach to collaboratively address physical health, behavioral health, and needs related to social determinants of health (SDoH)
- Utilization of weekly "gap in fill" reports to outreach members who have missed refilling their sickle cell disease modifying medications
- A partnership with the VCU Adult Sickle Cell Clinic that includes a monthly round meeting to bring together health plan medical directors, pharmacy team members, and Case Managers with VCU providers, patient navigators, and support staff to discuss member needs, successes, and barriers to care.
- A Life Skill Building program that connects SCD members with a behavioral health provider in their community. The Life Skill Building Program uses an integrated care approach to assist members with the development of skills needed to effectively manage their physical health condition and improve their quality of life.
    - assessment of the member's current skill set related to managing their physical health, navigating the healthcare system, managing their mental well-being,
        - includes assessment of social drivers impacting health
        - goals are established to measure the member's progress toward achieving successful life skills
    - member support system includes their primary Care Manager at the health plan, the Life Skill Building provider, the member's team of care providers, and any other identified member supports
    - may also include specialists, primary care providers, patient navigators, and therapists
    - goal of the Life Skill Building Program is to assist members with building skills that will allow them to have the best quality of life possible.
        - addressing basic and psychological needs through skill development will allow the member to better manage their physical and mental health

Early and Periodic Screening, Diagnostic, and Treatment

The Early and Periodic Screening, Diagnostic and Treatment program (EPSDT) provides global benefit global coverage for Medicaid members under the age of 21 who are living with SCD. EPSDT includes periodic screening, vision, dental and hearing services. EPSDT also includes a federal requirement which compels state Medicaid agencies to cover services, products, or procedures for children if those items are determined to be medically necessary to "correct or ameliorate" a defect, physical or mental illness, or health problem identified through routine medical screening or examination, regardless of whether coverage for the same

service/support is available to adults under the state plan.

Addressing Social Determinants of Health

The World Health Organization identifies social determinants of health (SdoH) as a key factor in determining the health status of individuals with chronic complications. Research indicates that SDoH, such as inadequate housing, food insecurity, and limited access to affordable healthcare, shape the health behavior of patients with SCD[iii]. Ninety-three percent of Virginia Medicaid's population diagnosesd with SCD is enrolled in managed care. Virginia Medicaid's managed care plans offer enhanced benefits which can help to address the social needs of Medicaid enrollees diagnosed with SCD such as providing resources for housing and food security as well as community supports.

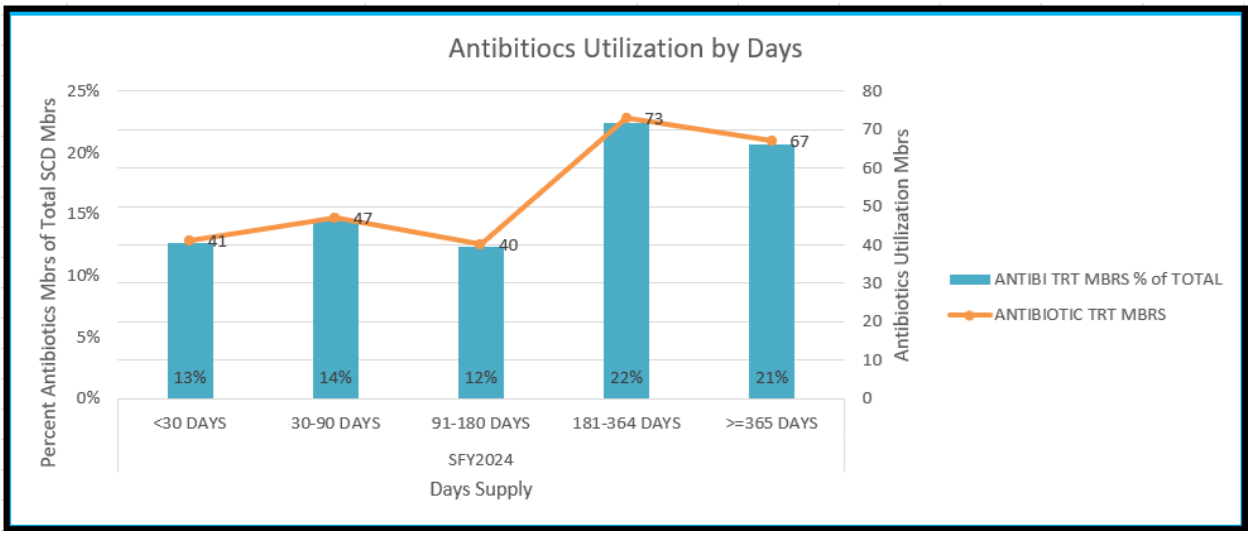
Utilization of Covered Services

Service category	% Medicaid Population diagnosed with SCD Utilizing Service
Primary Care	98%
Emergency Room	27%
Inpatient Hospital	36%
Pharmacy Services	87%

Medication Utilization in Eligible Medicaid and CHIP Enrollees with SCD			
Medication	**Number of Enrollees with Filled Prescription**	**Number Eligible for Medication**	**% Eligible Population w/Filled Prescription**
Hydroxyurea	779	2,331	33%
L-glutamine	37	2,006	2%
Voxelotor*	179	2,000	9%
Crizanlizumab	68	1,374	5%

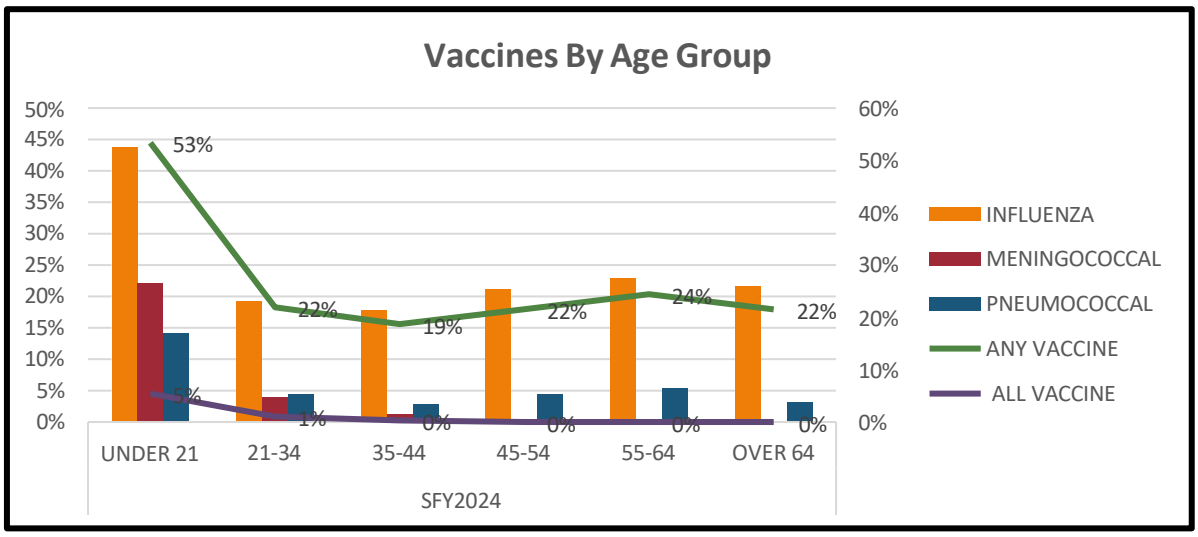
** Removed from the market Sep 2024. 179 Medicaid/CHIP members with SCD will be impacted by the removal of Voxelotor (Oxbryta) from the market.*

Figure 1: Utilization of Prophylactic Antibiotics- Medicaid and CHIP Members Under 6 Years of Age



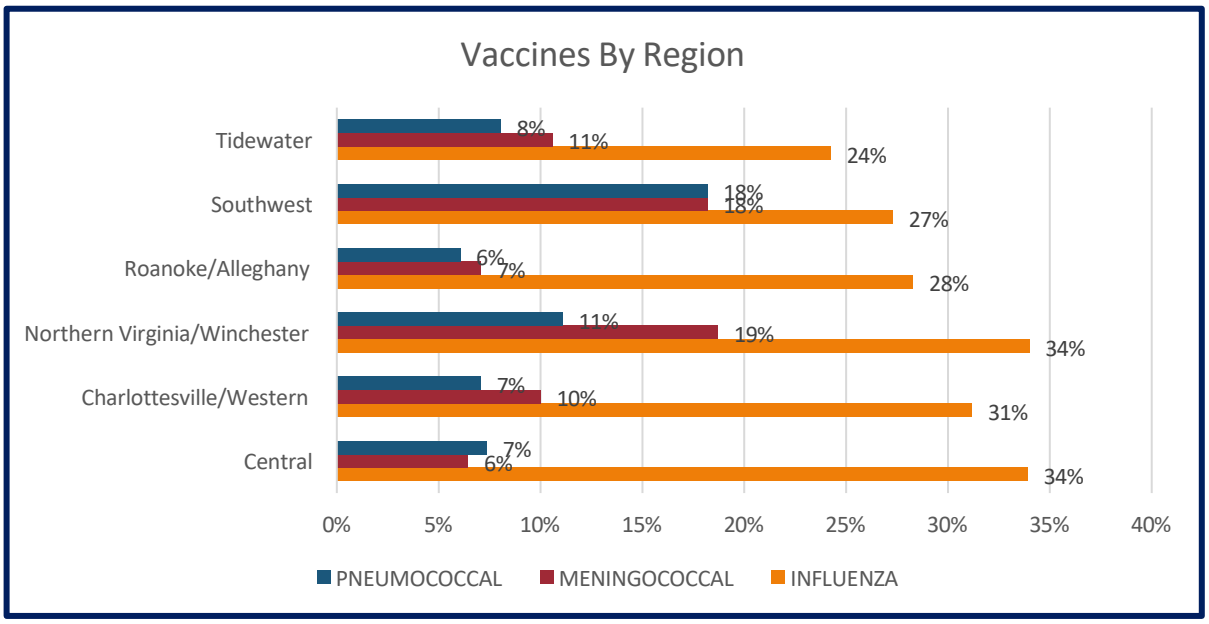
Children under 6 have been largely successful in adhering to a daily prophylactic antibiotic regimen with 21% of the population meeting 366 days

Figure 2: VA Medicaid and CHIP SCD Members Recommended Vaccinations by Age Group



Individuals living with SCD under 21 years of age received the highest percentage of recommended vaccinations. All ages are more likely to receive flu shots. Those aged 35-44 are less likely to receive any recommended vaccinations.

Figure 3: Percent of SCD Population Receiving Recommended Vaccinations by Region



The southwest region has the highest percentage of pneumococcal vaccinations with 18% of the SCD population receiving the vaccine. Central and Northern regions have the highest number of influenza vaccines with 34% of the SCD population receiving a flu shot. The Northern region has the highest percentage of SCD population receiving a meningococcal vaccine.

During SFY2024, 56% of children aged 5 and under received a flu vaccine and 30% received flu

vaccine across all age groups. 34% of Virginia Medicaid/CHIP enrollees aged 5 and under received a pneumococcal vaccine during SFY2024 and 21% received a daily antibiotic.

Summary

DMAS provides coverage for all SCD evidence-based treatments under Fee-for-Service (FFS) and managed care. Virginia Medicaid data shows gaps in recommended care for Medicaid and CHIP members with SCD, specifically in rates of transcranial Doppler ultrasound screening, pneumococcal vaccination for children, and hydroxyurea use among children and adults. DMAS provides coverage for SCD support services such as dental, vision, hearing, and transportation under FFS and managed care. Analysis of Medicaid data indicates that these services are under-utilized by the SCD population.

Transitioning from pediatric to adult SCD care can create barriers to treatment due to gaps in insurance coverage. Changes in Medicaid eligibility rules (e.g., extending Medicaid coverage to former foster care youth (2014), Medicaid Expansion coverage of childless adults (2019), and 12-month postpartum coverage extension for pregnant women (2021)) have had a positive impact on decreasing gaps in insurance coverage for Medicaid enrollees diagnosed with SCD who are transitioning from pediatric to adult care.

Barriers to treatment also exist due to a limited number of treatment centers. As of January 2023, the Virginia Department of Health (VDH) noted 9 Virginia Sickle Cell/ Hemoglobinopathy Treatment Centers. There are limited treatment centers for pediatric or adult SCD care in the far southwest. In the fall of 2024, Sentara Norfolk General Hospital and Eastern Virginia Medical School opened the Sentara-EVMS Comprehensive Sickle Cell Program which brings specialists from general internal medicine, hospital medicine, pain management, and psychiatry together in a single location to treat adults with SCD.

Area	Pediatric	Adult
Charlottesville	University of Virginia Medical Center	Emily Couric Clinical Cancer Center
Fairfax	Pediatric Specialists of Virginia	Inova Schar Cancer Institute
Norfolk	Children's Hospital of the King's Daughters	Sentara-EVMS Comprehensive Sickle Cell Program
Richmond	Children's Hospital of Richmond at VCU	VCU Health
Roanoke	Carilion Clinic Children's Hospital	Carilion Roanoke Memorial Hospital

Recommendations

- Utilize forums such as the Medicaid Member Advisory Committee, the Medicaid Physician and Managed Care Liaison Committee and the Medicaid Managed Care Advisory Committee to create opportunities for input from the SCD community (patients, families, community-based organizations, and providers) within DMAS forums
- DMAS and managed care partners pursue opportunities to collaborate on messaging to increase awareness of services

- Managed care partners continue to provide resources and tools relevant to disease education, medication adherence monitoring, and care coordination for healthcare providers who treat individuals living with SCD
- Continue partnership with Virginia Health Information (VHI) and managed care partners to increase awareness of SmartChart and the utilization of Care Insights to create individualized plans of care for individuals living with SCD
- DMAS Drug Utilization Review Board (DUR) to continue studies of underutilization of SCD disease-modifying treatments; DMAS DUR and the Pharmacy & Therapeutics Committee (P&T) continue to ensure access to prescription medications
- Foster data sharing agreements between VDH and DMAS concerning screening results and SCD/SCT status

About DMAS and Medicaid

The mission of the Virginia Medicaid agency is to improve the health and well-being of Virginians through access to high-quality health care coverage. The Department of Medical Assistance Services (DMAS) administers Virginia's Medicaid and CHIP programs for approximately two million Virginians. Members have access to primary and specialty health services, inpatient care, dental, behavioral health as well as addiction and recovery treatment services. In addition, Medicaid long-term services and supports enable thousands of Virginians to remain in their homes or to access residential and nursing home care.

Medicaid members historically have included children, pregnant women, parents and caretakers, older adults, and individuals with disabilities. In 2019, Virginia expanded the Medicaid eligibility rules to make health care coverage available to more than 600,000 newly eligible, low-income adults.

Medicaid and CHIP (known in Virginia as Family Access to Medical Insurance Security, or FAMIS) are jointly funded by Virginia and the federal government under Title XIX and Title XXI of the Social Security Act. Virginia generally receives an approximate dollar-for-dollar federal spending match in the Medicaid program. Medicaid expansion qualifies the Commonwealth for a federal funding match of no less than 90% for newly eligible adults, generating cost savings that benefit the overall state budget.

[i] *Source: American Society of Hematology 2019-2021 Clinical Practice Guidelines on Sickle Cell Disease.*
[ii] *Source: Mayo Clinic, Sickle Cell Anemia Diagnosis and Treatments*
[iii] *Source: Sickle Cell Disease and Social Determinants of Health- A Scoping Review*