

Specialty Guideline Management (SGM) Therapy and Drug Overview

ACROMEGALY

Acromegaly is a hormonal disorder caused by excessive production of growth hormone by the pituitary gland. It is a slowly progressive disease that mainly leads to soft tissue and bone overgrowth, which can result in severe disfigurement, serious complications and premature death.

octreotide (Sandostatin)	Somatuline Depot
Sandostatin LAR Depot	Somavert
Signifor LAR	

ALCOHOL AND OPIOID DEPENDENCY

Alcohol and opioid dependence can have severe impact on both physical and mental health. Vivitrol is an injectable medication used to treat alcohol and opioid dependence.

Vivitrol

ALLERGIC ASTHMA

Asthma is a chronic inflammatory disorder of the airways characterized by periods of reversible airway obstruction and airway inflammation. Asthma is frequently triggered by environmental allergens such as dust mites and pollens. Common symptoms include wheezing, coughing, chest tightness and shortness of breath. These symptoms may result in emergency room visits and hospitalization. People with allergic asthma often have specific immunoglobulin E (IgE) antibodies to allergens. Xolair inhibits the binding of IgE to the mast cells and basophils, decreasing the body's reaction to allergic stimuli.

Cinqair	Nucala
Fasenra	Xolair

ALPHA1-ANTITRYPSIN (AAT) DEFICIENCY

AAT deficiency is a hereditary disorder that increases the risk of lung disease (e.g., emphysema, chronic obstructive pulmonary disease), liver disease and other conditions. AAT is a serum protein that protects the lungs from enzymes of inflammatory cells, especially neutrophil elastase. AAT replacement products are used to treat AAT deficiency.

Aralast NP	Prolastin-C
Glassia	Zemaira

ANEMIA

Anemia is caused by a reduction of functional red blood cells (RBCs), increased destruction of RBCs or blood loss. This results in an inadequate supply of oxygen-rich blood that leads to weakness, fatigue, dizziness and shortness of breath. These medications increase the amount of RBCs, which increases the amount of oxygen-rich blood in the body.

Aranesp	Mircera
Epogen	Procrit

ATOPIC DERMATITIS

Atopic Dermatitis (AD) is a chronic, inflammatory skin condition characterized by a dry, red rash with itching. Dupixent is the first specialty drug approved for treatment of moderate to severe AD for patients whose disease is not adequately controlled by topical prescription therapies.

Dupixent

BONE DISORDERS—OTHER

Hypophosphatasia is a rare inherited disorder that affects the development of bones and teeth. It is caused by defective production of an enzyme that leads to the disruption of bone formation. This condition causes rickets, osteomalacia, fractures, and tooth loss. Strensiq is an enzyme replacement therapy that replaces the defective enzyme.

Strensiq

BOTULINUM TOXINS

Botulinum toxin is commonly used for treatment of cervical dystonia, spasticity, strabismus, blepharospasm and urinary incontinence. It is also used for prophylaxis of chronic migraine headaches. These medications work by blocking the affected nerves, which results in weakening or paralysis of the related muscles.

Botox	Myobloc
Dysport	Xeomin

CARDIAC DISORDERS

Tikosyn is an antiarrhythmic agent used for conversion to and maintenance of normal heart rhythm in people with atrial fibrillation or atrial flutter. It works by blocking the cardiac potassium ion channels.

Tikosyn

SGM Therapy and Drug Overview *continued*

CENTRAL PRECOCIOUS PUBERTY (CPP)

CPP is a condition that causes early onset of puberty in boys and girls due to early activation of hormones that are responsible for puberty. The underlying cause of this early release is unknown. Leuprolide, Lupron Depot-PED and Supprelin LA suppress the release of hormones that initiate puberty.

leuprolide

Supprelin LA

Lupron Depot-PED

CRYOPYRIN-ASSOCIATED PERIODIC SYNDROMES (CAPS)

CAPS are comprised of three rare genetic autoinflammatory syndromes: familial cold autoinflammatory syndrome (FCAS), Muckle-Wells syndrome (MWS) and neonatal-onset multisystem inflammatory disease/chronic infantile neurologic cutaneous and articular syndrome (NOMID/CINCA). Common symptoms among the three disorders include periodic fever, inflammation and recurrent urticaria-like skin rash. These interleukin-1 targeted therapies are used to reduce inflammation.

**Arcalyst
Ilaris**

Kineret

CUSHING'S SYNDROME

Cushing's syndrome is an endocrine disorder caused by overproduction of the hormone cortisol. Excess cortisol can result in weight gain, high blood pressure, depression and diabetes. Korlym works by blocking the action of cortisol. Korlym is indicated to control high blood glucose levels related to Cushing's syndrome when surgery was unsuccessful or surgery is not an option. Signifor is indicated for Cushing's disease when pituitary surgery is not an option or has not been curative.

Korlym

Signifor

CYSTIC FIBROSIS (CF)

CF is a hereditary disease of secretory glands, including the glands that make mucus and sweat. CF causes mucus to be thick and sticky, resulting in impaired mucus clearance of the airways and subsequent chronic pulmonary infections. Tobramycin inhalation solution, TOBI Podhaler, Bethkis, Kitabis Pak and Cayston are inhaled antibiotics used to treat the bacterial infections commonly found in people with CF. Pulmozyme is a mucolytic agent that decreases sputum viscosity and improves pulmonary function. Kalydeco and Orkambi are used for CF patients with certain genetic mutations.

Bethkis

Pulmozyme

Cayston

Symdeko

Kalydeco

TOBI Podhaler

Kitabis Pak

**tobramycin inhalation
solution (TOBI)**

Orkambi

ELECTROLYTE DISORDERS

Samsca helps to correct significantly low sodium levels due to conditions such as heart failure and certain hormone imbalances. Signs and symptoms include nausea, vomiting, confusion, fatigue and seizures. Low sodium levels should be corrected gradually to avoid severe neurologic damage and death.

Samsca

GASTROINTESTINAL DISORDERS—OTHER

Short bowel syndrome (SBS) causes poor absorption of nutrients that may occur when part of the small intestine is absent due to an acquired condition (e.g., surgery) or a congenital condition. Symptoms of SBS include weight loss, malnutrition, diarrhea and dehydration. Common reasons for SBS in adults include Crohn's disease, cancer and cancer treatment, trauma and other types of intestinal damage or blockage. Gattex is indicated for the treatment of patients with SBS who are dependent on intravenous nutritional support. Zorbtive is a growth hormone indicated for the treatment of SBS in patients receiving specialized nutritional support. Cholbam is a bile acid therapy indicated for the treatment of bile acid synthesis disorders.

Cholbam

Ocaliva

Gattex

Zorbtive

GOUT

Gout is a condition caused by high uric acid levels resulting in painful, swollen and tender joints that can lead to bone damage. Krystexxa is used for refractory chronic gout that cannot be controlled by standard gout medications. Krystexxa converts uric acid to a more water-soluble form that is more easily eliminated from the body.

Krystexxa

GROWTH HORMONE (GH) AND RELATED DISORDERS

GH is produced by the pituitary gland and promotes growth and regulates metabolism. GH deficiency can be caused by congenital or acquired conditions. Recombinant GH products are used to replace GH in people with GH deficiency. Increlex is used to replace insulin-like growth factor (IGF-1) in children with IGF-1 deficiency.

Genotropin

Nutropin AQ

Humatrope

Omnitrope

Increlex

Saizen

Norditropin

Zomacton

SGM Therapy and Drug Overview *continued*

HEMATOPOIETICS

Mozobil is used in combination with granulocyte colony-stimulating factor in people with lymphoma and myeloma who are undergoing stem cell collection and transplantation.

Mozobil

HEMOPHILIA AND RELATED BLEEDING DISORDERS

Hemophilia and von Willebrand disease (vWD) are inherited bleeding disorders. Hemophilia is caused by a deficiency of coagulation factor VIII (hemophilia A) or factor IX (hemophilia B). vWD is the most common inherited bleeding disorder and occurs when there is a defect in the quantity or quality of von Willebrand factor. These products replace the missing clotting factors to help prevent or treat spontaneous and traumatic bleeds and to recover from surgery.

Advate

Adynovate

Afstyla

Alphanate

AlphaNine SD

Alprolix

Bebulin VH

BeneFIX

Coagadex

Eloctate

Feiba NF

Feiba VH

Helixate FS

Hemlibra

Hemofil M

Humate-P

Idelvion

Ixinity

Koate-DVI

Kogenate FS

Kovlatry

Monoclote-P

Mononine

Novoeight

NovoSeven

Nuwiq

Obizur

Profilnine SD

Rebinyn

Recombinate

Rixubis

Stimate Nasal Spray

Vonvendi

Wilate

Xyntha

Ribavirin (ribavirin capsules/tablets, Copegus, Moderiba, Rebetol, Ribasphere, Ribasphere RibaPak, Ribatab)

HEREDITARY ANGIOEDEMA (HAE)

HAE is a rare, but potentially life-threatening disorder of the immune system. HAE is characterized by recurrent episodes of severe swelling in the skin and soft tissues. These episodes can occur in the throat, tongue, face, extremities or abdomen. Management of HAE includes short-term and long-term prophylaxis as well as treatment of acute attacks.

Berinert

Cinryze

Firazyr

Haegarda

Kalbitor

Ruconest

HORMONAL THERAPIES

Hormonal therapies are used to treat cancer and other medical conditions. Lupron is also used for infertility and central precocious puberty (CPP).

Aveed

Eligard

Firmagon

leuprolide

Lupaneta Pack

Lupron Depot

Natpara

Trelstar

Triptodur

Vantas

Zoladex

HUMAN IMMUNODEFICIENCY VIRUS (HIV)

HIV is the virus that causes acquired immune deficiency syndrome (AIDS). HIV attacks the immune system by destroying CD4 positive (CD4+) T cells, a type of white blood cell that is vital to fighting off infection. Egrifta is used to reduce the excess abdominal fat in HIV-infected patients with lipodystrophy. Serostim is used to increase lean body mass and body weight and improve endurance in HIV associated wasting or cachexia.

Egrifta

Serostim

IMMUNE THERAPIES

These medications replace immune globulin, a protein critical in fighting infection and providing immunity.

Bivigam

Carimune NF

Cuvitru

Flebogamma

GamaSTAN S/D

Gammagard

Gammaked

Gammaplex

Gamunex

Hizentra

HyQvia

Octagam

Privigen

Daklinza

Epclusa

Harvoni

Mavyret

Olysio

Pegasys

Sovaldi

Technivie

Viekira Pak/Viekira XR

Vosevi

Zepatier

SGM Therapy and Drug Overview *continued*

IMMUNE (IDIOPATHIC) THROMBOCYTOPENIA (ITP)

ITP is an autoimmune disorder in which there are too few platelets due to an increased destruction and an inappropriately low production rate of new platelets. ITP increases the risk of serious and potentially life-threatening bleeding episodes. Nplate and Promacta are used to increase platelet production and reduce the risk for serious bleeding episodes.

Nplate

Promacta

INFECTIOUS DISEASE

Actimmune is used to treat chronic granulomatous disease (CGD), an inherited immunodeficiency disorder that makes a person vulnerable to various infections. Actimmune reduces the frequency and severity of the serious infections associated with CGD.

Actimmune

INFERTILITY

Infertility is medically defined as an inability to achieve pregnancy after 12 months or more of trying to conceive. Infertility medications can be used in assisted reproductive technology procedures, such as in vitro fertilization, or in ovulation induction procedures to help achieve pregnancy.

Bravelle

Cetrotide

chorionic

gonadotropin

(Novarel, Ovidrel,

Pregnyl)

Follistim AQ

ganirelix acetate

Gonal-F

leuprolide

Menopur

Repronex

INFLAMMATORY BOWEL DISEASE (IBD)

IBD covers a number of disorders where the digestive tract becomes inflamed due to abnormal actions by the immune system. The inflammation can result in pain, diarrhea and unintended weight loss. These medications help to reduce the inflammation to relieve symptoms and prevent progressive damage to the digestive tract.

Cimzia

Entyvio

Humira

Inflectra

Remicade

Renflexis

Simponi

Stelara

Tysabri

IRON OVERLOAD

Iron is an important component of hemoglobin, the chemical in red blood cells that transports oxygen. Some people accumulate excessive amounts of iron due to genetic disorders or through repeat blood transfusions. The excess iron is deposited in the liver and other organs where it can interfere with the organ's function. These medications assist in the removal of iron from the body.

deferoxamine (Desferal)

Exjade

Ferriprox

Jadenu

LIPID DISORDERS

Familial hypercholesterolemia (FH) is an inherited lipid disorder that causes high cholesterol levels and an early onset of cardiovascular disease. Homozygous FH is a rare, but severe, form of FH that is associated with very high levels of cholesterol, cholesterol deposits in the tendons, skin, or other areas, severe heart disease at a young age and a poor response to standard lipid-lowering therapy. Juxtapid and Kynamro help reduce cholesterol levels in people with homozygous.

Juxtapid

Kynamro

Praluent

Repatha

LIPODYSTROPHY

Lipodystrophy is a rare disorder caused by abnormalities in fat tissue distribution and a loss of the fat tissue necessary for normal metabolism. Lipodystrophy may lead to complications such as diabetes and lipid disorders. Myalept is used with dietary changes to treat lipodystrophy.

Myalept

LYSOSOMAL STORAGE DISORDERS (LSD) AND RELATED DISORDERS

Lysosomes contain enzymes within cells that digest large biomolecules into smaller usable fragments. LSDs are genetic disorders caused by the lack of an enzyme that breaks down these large molecules. Over time, the build-up of the undigested substance damages the cells and thus the functioning of the organ to which they belong. These medications treat LSDs by replacing the missing enzyme.

Adagen

Aldurazyme

Brineura*

Cerdelga

Cerezyme

Cystagon

Cystaran

Elaprase

Elelyso

Fabrazyme

Kanuma

Lumizyme

Mepsevii

Naglazyme

Nityr

Orfadin

Procysbi

Vimizim

VPRIV

Zavesca

MOVEMENT DISORDERS

Apokyn helps treat people with Parkinson's disease who have difficulty moving. Xenazine helps treat the involuntary spasms or jerky movements associated with Huntington's disease. Northera helps treat drops in blood pressure associated with standing too quickly in people with underlying neurological conditions.

SGM Therapy and Drug Overview *continued*

Apokyn
Austedo
Ingrezza
Northera

Nuplazid
Radicava
Xenazine

Bavencio
Beleodaq
Bendeka
Besponsa
Blincyto
Bosulif
Cabometyx
Calquence
capecitabine (Xeloda)

Caprelsa
Cometriq
Cotellic
Cyramza
Darzalex
decitabine (Dacogen)

Empliciti
Erbitux
Erivedge
Erleada
Erwinaze

Farydak
Folotyn
Fusilev
Gazyva
Gilotrif
Gleevec

Halaven
Herceptin
Hycamtin Capsules

Ibrance
Iclusig
Idhifa
Imbruvica
Imfinzi
Imlygic

Inlyta
Intron-A
Iressa
Istodax
Ixempra
Jakafi

Jevtana
Kadcyla
Keytruda
Kisqali/Kisqali Pak Femara
Kymriah*

Kyprolis
Lartruvo
Lenvima
Lonsurf
Lynparza
Mekinist
mitoxantrone

Mylotarg
Nerlynx
Nexavar
Ninlaro
Odomzo

Oncaspar
Opdivo
Perjeta
Pomalyst
Portrazza
Proleukin
Revlimid

Rituxan
Rituxan Hycela
Rubraca
Rydapt
Sprycel
Stivarga

Sutent
Sylatron
Sylvant
Synribo
Tafinlar
Tagrisso

Tarceva
Targretin
Tasigna
Tecentriq
temozolomide (Temodar)

Thalomid
Torisel
Treanda
Tykerb
Valchlor
Vectibix

Velcade
Venclexta
Verzenio
Votrient
Xalkori
Xermelo

Xgeva
Xtandi
Yervoy
Yescarta*
Zaltrap
Zejula

Zelboraf
zoledronic acid (Zometa)
Zolinza
Zydelig
Zykadia
Zytiga

MULTIPLE SCLEROSIS (MS)

MS is characterized by damage to nerves in the brain and spinal cord that prevents those nerves from communicating. When the nerves cannot communicate, people begin to lose control of muscle movements and other functions. Ampyra helps improve walking ability in people with MS. The other medications help prevent progression of the disease to limit the loss of function.

Ampyra

Lemtrada

Aubagio

mitoxantrone

Avonex

Ocrevus

Betaseron

Plegridy

Copaxone

Rebif

Extavia

Tecfidera

Gilenya

Tysabri

Glatopa

Zinbryta

MUSCULAR DYSTROPHY

Exondys 51 is a treatment for Duchenne Muscular Dystrophy (DMD). DMD is a rare, inherited muscle disease.

Emflaza

Exondys 51

NEUROMUSCULAR

Spinraza™ is a treatment for spinal muscular atrophy (SMA) This specialty drug is administered intrathecally under sedation, and will most commonly be administered in a hospital or surgical center.

Spinraza*

NEUTROPENIA

Neutropenia is an abnormally low count of neutrophils, a type of white blood cell that protects the body from infection. The reduced number of neutrophils may be due to a number of causes, but it is a common adverse effect of cancer chemotherapy. These medications stimulate the production of more white blood cells.

Granix

Neupogen

Leukine

Zarxio

Neulasta

ONCOLOGY

These medications are used to treat various cancers. Each agent targets different tumor types.

Adcetris

Alunbrig

Afinitor

Arzerra

Alecensa

Avastin

Aliqopa

azacitidine (Vidaza)

SGM Therapy and Drug Overview *continued*

OSTEOARTHRITIS (OA)

OA is a common form of arthritis that causes pain, swelling and reduced motion in the affected joints. In people with knee osteoarthritis, the joint fluid breaks down and does not provide adequate cushioning to the joint. These medications supplement the joint fluid to help reduce pain and improve joint function.

Euflexxa	Hymovis
Durolane	Monovisc
Gel-One	Orthovisc
Gelsyn-3	Supartz
Genvisc-850	Synvisc/Synvisc One
Hyalgan	Visco-3

OSTEOPOROSIS

Osteoporosis is characterized by reduced bone density and an increased risk of fracture. Bone fractures cause significant pain and may require hospitalization or surgery. These medications increase or stabilize bone density to reduce fracture risk.

Forteo	Tymlos
Prolia	zoledronic acid (Reclast)

PAROXYSMAL NOCTURNAL HEMOGLOBINURIA (PNH)

PNH is a rare disorder where the immune system attacks blood cells. The destruction of red blood cells can cause anemia and blood clots. Soliris blocks the immune system from attacking the blood cells which helps prevent anemia and blood clots.

Soliris

PHENYLKETONURIA (PKU)

PKU is a rare genetic disorder that impairs the body's ability to metabolize phenylalanine (Phe), an amino acid found in protein. As Phe accumulates in the body, it can interfere with brain function, causing impaired thinking and behavioral problems. Kuvan helps the body metabolize Phe to reduce and prevent the negative effects of Phe accumulation.

Kuvan

PRE-TERM BIRTH

Makena is used to reduce the risk of pre-term birth in women who have a history of delivering a baby too early.

Makena

PSORIASIS

Psoriasis is a chronic, autoimmune skin disease that causes itchy or sore patches of thick, red skin with silvery scales known as plaques. Severe cases can be painful, disfiguring and disabling. These medications help prevent the immune system from attacking the skin.

Cosentyx	Remicade
Enbrel	Renflexis
Humira	Siliq
Inflectra	Stelara
Otezla	Taltz
Otrexup	Tremfya
Rasuvo	

PULMONARY ARTERIAL HYPERTENSION (PAH)

PAH is a condition characterized by high blood pressure in the lungs, which puts strain on the heart. Symptoms can include shortness of breath, dizziness and decreased exercise tolerance. People with severe disease have difficulty breathing, even at rest. PAH medications help improve symptoms.

Adcirca	sildenafil (Revatio)
Adempas	Tracleer
epoprostenol (Flolan)	Tyvaso
Letairis	Uptravi
Opsumit	Veletri
Orenitram	Ventavis
Remodulin	

PULMONARY DISORDERS—OTHER

Idiopathic pulmonary fibrosis is chronic, progressive and ultimately fatal scarring of the lung tissue due to an unknown cause. As the lung tissue becomes more scarred, oxygen cannot move into the body as efficiently and the lungs become less flexible. These medications help stabilize the patient's lung function and slow the rate of lung function decline.

Esbriet	Ofev
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RARE DISORDERS—OTHER

The definition of a rare disease includes a disease that affects fewer than 200,000 people in the United States. Drugs included in the Rare Disorder—other category include products that treat conditions which meet the definition of a rare condition, have only 1 or 2 drugs available for the treatment of that condition, and treat a condition that cannot be classified under any other category in the SGM Therapy Drug List.

Luxturna* (gene therapy for vision loss)

SGM Therapy and Drug Overview *continued*

RENAL DISORDERS

People with chronic kidney disease can have low calcium levels. The body tries to correct this imbalance by producing more parathyroid hormone, which stimulates the breakdown of bone and increases calcium levels. Sensipar helps regulate calcium levels in people with kidney disease or with other causes of parathyroid hormone imbalance.

Parsabiv

Sensipar

RESPIRATORY SYNCYTIAL VIRUS (RSV)

RSV is a virus that causes severe cold-like symptoms, which may lead to emergency department visits and hospitalizations. Infants born prematurely and those born with lung and heart disease are at high risk for RSV infection. Synagis is used to prevent RSV infection in high-risk infants.

Synagis

RETINAL DISORDERS

Macular degeneration occurs when there is a deterioration of the retina that leads to vision loss. These medications help slow the progression of the disease.

Avastin
Eylea

Lucentis

RHEUMATOID ARTHRITIS (RA)

RA is an autoimmune disease that causes pain, swelling and stiffness in joints. If untreated, this swelling may lead to joint deformity and disability. These medications are disease-modifying agents that act on the immune system to slow the progression of RA.

Actemra
Cimzia
Enbrel
Humira
Inflectra
Kevzara
Kineret
Orencia

Otrexup
Rasuvo
Remicade
Renflexis
Rituxan
Simponi
Simponi Aria
Xeljanz/ Xeljanz XR

SEIZURE DISORDERS

Seizure disorders are disruptive to a person's life and, if uncontrolled, can cause severe problems such as injury to the body or brain, decreased quality of life and intellectual decline in children. These medications help prevent seizures in people with specific types of seizure disorders.

Acthar

Sabril

SICKLE CELL DISEASE

Sickle cell disease is an inherited blood disorder characterized by abnormal hemoglobin (the oxygen-carrying protein within the red blood cells). The abnormal hemoglobin causes distorted (crescent shaped or "sickled") red blood cells. These abnormally shaped cells restrict the flow of blood and limit oxygen delivery which can lead to severe pain and organ damage.

Endari

SLEEP DISORDERS

Non-24-hour sleep-wake disorder (N24SWD) is a chronic circadian rhythm disorder that occurs when sleep and wake cycles are not synchronized with the normal 24-hour cycle. This condition is most commonly found in people who are totally blind with no perception of light. People with N24SWD cannot synchronize their internal clock with the external 24-hour environment. They often have difficulty sleeping at night and staying awake during the day. Hetlioz helps treat people with N24SWD.

Hetlioz

SYSTEMIC LUPUS ERYTHEMATOSUS (SLE)

SLE is a systemic autoimmune disease in which antibodies are produced against a person's own tissues. The tissues affected can be the skin, joints, kidneys and other organs. The antibodies and resulting immune response can cause swelling, pain and tissue damage. Benlysta helps reduce disease activity in SLE.

Benlysta

UREA CYCLE DISORDERS (UCDs)

UCDs are genetic disorders caused by a deficiency of one of the enzymes that remove ammonia from the bloodstream. When an enzyme in this process is deficient, ammonia or other urea precursors can accumulate. Accumulation of the ammonia can lead to seizures, respiratory distress and possibly death. These medications help reduce the ammonia level in the blood.

Buphenyl
Carbaglu

Ravicti

SGM Therapy and Drug Overview *continued*



With more than forty years of specialty pharmacy experience, CVS Specialty® provides proactive quality care and service. We have a network of pharmacies that includes those with Joint Commission and URAC accreditation. The Joint Commission and URAC are nationally-recognized symbols of quality which reflects an organization's commitment to meet high standards of quality and safety.

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