Carta Trámite

20 de enero de 2023

A: Todos los proveedores Contratados por First Medical Health Plan, Inc. para el Plan Vital, Región Única.

Re: ***Carta Normativa 23-0117-02 Inclusión de Albinismo Oculocutáneo, Síndrome de Hermansky-Pudlak (HPS) y Síndrome de Chediak-Higashi (CHS) en la Cubierta Especial y Política de Manejo de los Pacientes Diagnosticados con esta Condición***

Estimado(a) Proveedor(a):

Reciba un cordial saludo de parte de First Medical Health Plan, Inc., (FMHP).

Adjunto a este comunicado encontrará la Carta Normativa 23-0117-02 de la Administración de Seguros de Salud (ASES).

A través de esta Carta Normativa, la ASES informa la implementación inmediata de la Política **ASES-OC-2023/P004**, con el propósito de uniformar el acceso a cuidados preventivos, diagnósticos y terapéuticos que necesitan los beneficiarios diagnosticados con **Albinismo Oculocutáneo, Síndrome de Hermansky-Pudlak (HPS) y Síndrome de Chediak-Higashi (CHS)**. Esta Política tiene efectividad inmediata.

Para detalles específicos, sobre la información provista por la ASES, le exhortamos a leer detenidamente Carta Normativa 23-0117-02 Inclusión de Albinismo Oculocutáneo, Síndrome de Hermansky-Pudlak (HPS) y Síndrome de Chediak-Higashi (CHS) en la Cubierta Especial y la ASES-OC-2023/P004.

Si usted tiene alguna pregunta relacionada a este comunicado y/o necesita información adicional, siéntase en la libertad de comunicarse con nuestro Centro de Servicio al Proveedor al número libre de cargos 1-844-347-7802 de lunes a viernes de 7:00 a.m. a 7:00 p.m. También, puede acceder a nuestra página electrónica www.firstmedicalvital.com.

Cordialmente,

Departamento de Cumplimiento
First Medical Health Plan, Inc.

GOBIERNO DE PUERTO RICO
ADMINISTRACIÓN DE SEGUROS DE SALUD
Directora Ejecutiva | Edna Y. Marín Ramos | emarin@asespr.org

Carta Normativa 23-0117-02

17 de enero de 2023

A: Organizaciones contratadas de Manejo Coordinado de Salud (MCO), Grupos Médicos Primarios (GMP), y Proveedores Participantes del Plan Vital

RE: INCLUSIÓN DE ALBINISMO OCULOCUTÁNEO, SÍNDROME DE HERMANSKY-PUDLAK (HSP) Y SÍNDROME DE CHEDIA-HIGASHI (CHS) EN LA CUBIERTA ESPECIAL Y POLÍTICA DE MANEJO DE LOS PACIENTES DIAGNOSTICADOS CON ESTA CONDICIÓN

Se adjunta la política # **ASES-OC-2022/P004,** la cual procura uniformar el acceso a cuidados preventivos, diagnósticos y terapéuticos que necesitan los beneficiarios diagnosticados con **Albinismo Oculocutáneo, Síndrome de Hermansky-Pudlak, y Síndrome de Chediak-Higashi (CHS)**.

La política aneja tiene efectividad de inmediato. Por lo tanto, requerimos a todas las aseguradoras contratadas bajo Plan Vital que se aseguren de cumplir con lo establecido en la misma. Igualmente, les solicitamos que se aseguren de diseminar el contenido de la política entre sus respectivas redes de proveedores contratados.

Cordialmente,

Roxanna K. Rosario Serrano, BHE, MS
Subdirectora Ejecutiva

Anejo (1)

Planificación, Calidad y Asuntos Clínicos **Plan de salud del Gobierno (PSG)** **SALUD VITAL**		
Policy: **Albinismo Oculocutáneo, Síndrome de Hermansky-Pudlak (HPS) y Síndrome de Chediak-Higashi (CHS).**		
Number: **ASES-OC-2023/P004**	**Effective Date:** **January 1, 2023**	**Number of Pages:** 11
Approved By: Roxanna K. Rosario Serrano, BHE, MS Deputy Executive Director	**Signature:**	**Date:** January 17, 2023
Reference: Contract Section ATTACHMENT 7		

PROPÓSITO

Establecer un protocolo uniforme para incluir bajo la cubierta especial de VITAL los casos de Albinismo Oculocutáneo, Síndrome de Hermansky-Pudlak, y síndrome de Chediak-Higashi (CHS) para uniformar el acceso a cuidados preventivos, diagnósticos y terapéuticos necesarios para el manejo adecuado, ágil y eficaz.

INTRODUCCIÓN

El albinismo es una condición genética poco frecuente asociada fundamentalmente a una deficiencia visual importante, que es específica y característica. Adicionalmente, la mayoría de las personas con albinismo pueden manifestar una pérdida parcial o total ausencia de pigmento, que sin embargo no aparece en todos los casos. Hay muchos tipos de albinismo. Conocemos por lo menos 20 genes cuyas mutaciones están asociadas a algún tipo de albinismo. Podemos conocer la causa del albinismo mediante diagnóstico genético. No existen todavía terapias aprobadas que puedan administrarse a personas con albinismo. Pero sí que hay diversas propuestas experimentales que se están investigando en la actualidad.

El albinismo es una condición hereditaria que se presenta al nacer y se caracteriza esencialmente por una ausencia o disminución del pigmento que da color a la piel, cabello y ojos. Se calcula que uno de cada 17,000 personas tiene algún tipo o variedad de albinismo. Afecta o se manifiesta en todas las razas.

En algunos casos las manifestaciones de albinismo son tan sutiles que ni la propia persona sabe que lo padece.

De una manera muy general, se pude considerar que hay dos grandes grupos de albinos:

1 Albinismo Oculocutáneo (OCA) por sus siglas en inglés, y
2 Albinismo Ocular (OA).

En el primer caso u OCA se describe una disminución de pigmento en los ojos, cabello y piel.
De este subgrupo de han descrito siete tipos o variantes, dependiendo del defecto genético que lo ocasiona. En el caso de albinismo ocular, solo suele afectarse los ojos, mientras que la piel y el cabello pudiera ser normal o presentar una coloración casi normal.

El albinismo se hereda de manera autosómica recesiva, por lo que para que se manifieste, tanto el padre como la madre deben tener el gen, y en esos casos existe un 25% de probabilidad de que cada embarazo sea de albino. En muchos casos los padres no tienen manifestaciones de albinismo, pero son portadores del gen del albinismo.

En el caso de OA, el gen del albinismo se encuentra localizado en el cromosoma X.
Este tipo de albinismo se manifiesta casi exclusivamente en varones y es transmitido por una madre portadora. En estos casos existe un 50% de probabilidades de tener un hijo con albinismo ocular (OA).

VARIANTES DE OCA:
No todas las personas con albinismo oculocutáneo presentan las mismas características.

OCA1: Es la variante más frecuente y corresponde al 43% de todos los OCA. Es el más frecuente en poblaciones occidentales (América y Europa). Es el tipo característico de albinismo en el que

todo el mundo piensa cuando le preguntan por el aspecto de una persona con albinismo. Es decir, una persona de pelo blanco, piel blanca o rosada muy pálida, con ojos muy claros o rojos/rosados. El albinismo OCA1 está causado por mutaciones o alteraciones en el gen de la tirosinasa (*TYR*, situado en el cromosoma 11 humano), que lleva la información genética de una de las principales enzimas responsable del primer paso de la ruta de síntesis de la melanina.

La mayoría de las personas con albinismo OCA1 presentan una agudeza visual alrededor del 10-20%, manifiestan nistagmo, fotofobia y visión tridimensional reducida.

OCA2: Mas frecuentes en personas de origen africano. El albinismo oculocutáneo de tipo 2 (OCA2), es el tipo más frecuente de albinismo en personas de raza negra, de origen africano. Está producido por mutaciones o alteraciones en el gen OCA2 (situado en el cromosoma 15 humano). Las personas con albinismo OCA2 suelen tener una agudeza visual entre el 20% (0,2) y el 30% (0,3). El color de la piel es blanco cremoso mientras que el color del pelo suele ser entre amarillo y marrón claros. Pueden aparecer pelos más oscuros en otras partes del cuerpo. El color de los ojos suele ser entre azul y marrón.

OCA3: El albinismo oculocutáneo de tipo 3 (OCA3) se asocia a mutaciones o alteraciones del gen de la proteína relacionada con tirosinasa de tipo 1 (*TYRP1*), otra de las enzimas componentes de la síntesis de eumelanina. Es un tipo de albinismo oculocutáneo no sindrómico relativamente raro fuera de África. Las personas OCA3 BOCA tienen un color de pelo entre claro y marrón y un color de piel igualmente de claro a marrón o tostado. En cuanto a las alteraciones visuales, el OCA3 probablemente sea el tipo de albinismo más leve, con una agudeza visual mayor y, frecuentemente, sin aparición de nistagmo.

OCA4: El albinismo oculocutáneo de tipo 4 (OCA4) está asociado a mutaciones o alteraciones en el gen *SLC45A2*, que codifica para una proteína de transporte asociada a melanosomas. Es el tipo de albinismo más frecuente en Japón. El color de la piel de las personas con albinismo OCA4 suele ser blanco cremoso. El color del pelo oscila entre blanco plateado y amarillo claro, y puede oscurecerse con el tiempo. El color de los ojos varía entre azul y marrón. Las personas con albinismo OCA4 pueden presentar graves deficiencias visuales, con agudezas visuales del 10% o

valores inferiores. También presentan nistagmo y visión estereoscópica reducida. No es infrecuente el estrabismo.

OCA5: El albinismo oculocutáneo de tipo 5 (OCA5) corresponde a mutaciones o alteraciones en un gen, todavía desconocido, asociado a la región cromosómica *4q24* del genoma humano. Ha sido detectado por vez primera en familias de origen paquistaní. La descripción de los pocos casos todavía detectados de personas con albinismo OCA5 recuerda a otros tipos de albinismo: piel blanca, pelo de color dorado, nistagmo, fotofobia, hipoplasia de la fóvea (ausencia de fóvea) y agudeza visual reducida.

OCA6: El albinismo oculocutáneo de tipo 6 (OCA6) es, junto al OCA7, uno de los últimos tipos de albinismo descritos y es extraordinariamente raro. Los primeros casos de OCA6 fueron descritos por vez primera en familias de origen chino, pero actualmente personas con albinismo OCA6 ya han sido detectadas también en Europa. Las personas con albinismo OCA6 tienen la piel blanca, color de cabellos claro (no blanco), que puede oscurecerse con el tiempo, hipoplasia de la fóvea, nistagmo, fotofobia, iris transparente y agudeza visual limitada.

OCA7: El albinismo oculocutáneo de tipo 7 (OCA7) corresponde a mutaciones o alteraciones del gen *LRMDA*, que codifica para una proteína involucrada en la diferenciación de los melanocitos. Este nuevo tipo de albinismo fue detectado por vez primera en familias danesas de las islas Feroe y en una persona de Lituania. Las personas con albinismo OCA7 tienen la piel blanca, más clara que la de sus padres. El color de pelo puede presentarse entre rubio claro a marrón oscuro. Presentan nistagmo, iris transparente y una agudeza visual limitada, que oscila entre un 5% (0,05) y un 30% (0,3).

Los arriba mencionados se catalogan como albinismos no sindrómicos, para diferenciarlos de otros albinos con condiciones asociadas como HERMANSKY-PUDLAK y el CHEDIAK-HIGASHI, que se denominan albinismos sindrómicos.

SÍNDROME DE HERMANSKY-PUDLAK (HPS)

Además de los anteriores, existen otros tipos de albinismo oculocutáneo, mucho menos frecuentes, en los que la disminución o ausencia de pigmento en piel, pelo y ojos se manifiesta

de forma combinada con otros síntomas, dentro de síndromes más complejos. Es el caso del síndrome de Hermansky-Pudlak (HPS), del cual existen 10 variantes, muy raro en la población en general, con una prevalencia que oscila entre 1:500.000 y 1:1.000.000 (con excepción de Puerto Rico, en donde, debido a un efecto fundador, el subtipo HPS1 representa el tipo de albinismo más común, detectado en aproximadamente 1 de cada 1.800 personas).

El efecto "fundador" se explica por la aparición espontánea de una mutación en un gen en una población en un momento determinado de la historia. Debido a la geografía del lugar, que puede limitar los movimientos de la población (por ejemplo, el convivir en una isla) y a determinados porcentajes de consanguinidad (las parejas se establecen entre individuos genéticamente relacionados) acaba expandiéndose y aparece en muchos individuos de esa misma población.
El HPS, además de las características propias del albinismo oculocutáneo (problemas de pigmentación y de visión) se manifiesta adicionalmente con:

- **Manifestaciones dermatológicas:** queratosis solar, carcinoma de células escamosas y basocelulares, nevos melanocíticos, acantosis nigricans en cuello y axila, hipertricosis y múltiples equimosis.
- **Trastornos hematológicos** como disfunción plaquetaria y diátesis hemorrágicas.
- **Problemas respiratorios** debidos a fibrosis pulmonar, el problema más grave y discapacitante, desgraciadamente con consecuencias fatales y principal causa de mortalidad en HPS1, desarrollándola prácticamente el 100% de este grupo de pacientes. La mortalidad por esta condición suele ocurrir entre los 40-50 años.
- **Problemas gastroenterológicos** conocida como colitis asociada a HPS, con una presentación muy parecida a la enfermedad de Crohn.
- **Problemas oftalmológicos**: albinismo ocular, nistagmo periódico alternante, estrabismo, pobre visión, cataratas tempranas hipoplasia de la fóvea y otras lo que hace que la mayoría de estas personas sean no videntes legales.
- **Problemas renales:** Algunos pacientes desarrollan enfermedad renal crónica. La aparición de hemorragias frecuentes y/o hematomas subcutáneos en niños con albinismo oculocutáneo debe hacer sospechar de un posible diagnóstico de HPS o CHS.

SÍNDROME DE CHEDIAK-HIGASHI (CHS)

El síndrome de Chediak-Higashi (CHS) cursa con síntomas parcialmente similares a HPS (albinismo oculocutáneo, hemorragias, hematomas…), aunque presenta adicionalmente problemas muy graves del sistema inmunológico, con una susceptibilidad aumentada frente a infecciones. Como en los casos de HPS1 y HPS4, el CHS también puede ser mortal. Las complicaciones clínicas de CHS incluyen hepatoesplenomegalia (hígado y bazo inflamados, agrandados), hipertrofia de ganglios, infecciones recurrentes respiratorias y cutáneas piogénicas (que producen pus), resultado del déficit de células polinucleares y de los linfocitos NK (del inglés "*Natural Killer*") del sistema inmune. El pronóstico suele ser muy poco favorable, complicado frecuentemente con un deterioro neurológico progresivo. El único tratamiento actualmente es el trasplante de médula ósea. Sin el trasplante los pacientes tienen una supervivencia media de 3 años. Tras el trasplante no todos los pacientes consiguen sobrevivir.

HALLAZGOS CLÍNICOS GENERALES

• **Nistagmos:** Movimiento involuntario del globo ocular.

• P**ostura anómala de la cabeza**: El niño suele adoptar posturas no usuales para poder reducir el movimiento involuntario de los ojos y optimizar su visión.

• **Estrabismo**: desalineación del eje ocular.

• **Fotofobia:** sensibilidad aumentada a la luz y claridad

• Trastornos de refracción: Es común que presenten hipermetropía (hyperopia), miopía y Astigmatismo

• **Hipoplasia de la fóvea:** Desarrollo anormal de la parte central de la retina, esto ocasiona visión defectuosa o disminuida.

• **Alteración en la trayectoria del nervio óptico**: El nervio óptico en su paso de la retina al cerebro sigue una trayectoria anómala.

- El iris carece de pigmentación o esta disminuida, por lo que no protege adecuadamente al ojo de los rayos lumínicos, lo que se conoce como transiluminación del iris.
- La agudeza visual puede variar y estar cerca de lo normal o hasta tener ceguera legal (menos de 20/200) o incluso peor en los casos más severos de albinismo. La visión cercana

suele ser algo mejor que la visión a distancia. Generalmente, a menor cantidad de pigmento peor visión.

En los casos de HPS, El HPS, además de las características propias del albinismo oculocutáneo (problemas de pigmentación y de visión) se manifiesta adicionalmente con:

- **Manifestaciones dermatológicas**, queratosis solar, carcinoma de células escamosas y basocelulares, nevos melanocíticos, acantosis nigricans en cuello y axila, hipertricosis y múltiples equimosis.
- **Trastornos hematológicos** como disfunción plaquetaria y diátesis hemorrágicas,problemas respiratorios debidos a fibrosis pulmonar, el problema más grave y discapacitante, desgraciadamente con consecuencias fatales y principal causa de mortalidad en HPS1, desarrollándola prácticamente el 100% de este grupo de pacientes. La mortalidad por esta condición suele ocurrir entre los 40-50 años.
- **Problemas gastroenterológicos** conocida como colitis asociada a HPS, con una presentación muy parecida a la enfermedad de Crohn.
- **Problemas oftalmológicos**: albinismo ocular, nistagmo periódico alternante, estrabismo, pobre visión, cataratas tempranas hipoplasia de la fóvea y otras lo que hace que la mayoría de estas personas sean no videntes legales.
- **Problemas renales:** Algunos pacientes desarrollan enfermedad renal crónica.

PRUEBAS Y EXÁMENES DIAGNÓSTICOS

1- Las pruebas genéticas ofrecen la forma más precisa de diagnosticar el albinismo. Dichas pruebas son útiles si usted tiene antecedentes familiares de albinismo. También resultan útiles para ciertos grupos de personas que se sabe padecen esta enfermedad.

2- Electrorretinografía. Este es un examen que puede revelar problemas visuales relacionados con el albinismo. Es realizado por un oftalmólogo.
3- Potenciales visuales evocados. Es un estudio que puede ser muy útil cuando el diagnóstico es incierto.

4- Examen físico: Se puede diagnosticar la afección con base en la apariencia de la piel, el cabello y los ojos.

-Falta de color en el cabello, la piel o el iris del ojo

-Piel y cabello más claros de lo normal

-Parches de piel sin color

- Estrabismo

-Sensibilidad a la luz

-Movimientos oculares rápido

-Problemas de visión o no visión funcional y legal.

5- Otros exámenes oftalmológicos:

-Prueba de músculos visuales;

-prueba de agudeza visual;

-Pruebas de refracción;

-Campos visuales;

-Pruebas de colores;

-Examen con lámparas de hendidura;

-Tonometría;

-Examen de retina con dilatación pupilar

CRITERIOS DE INCLUSIÓN PARA CONDICIÓN ESPECIAL

Se requerirá una certificación diagnostica por uno de los siguientes especialistas:

Dermatólogo

Oftalmólogo

Genetista

Y

Resultados de exámenes o pruebas que sustenten el diagnóstico como:

- Pruebas genéticas
- Pigmentos de piel

- Estudios oftalmológicos

En el caso de OCA sindrómicos, también debe de acompañarse de una certificación por un hematólogo.

EFECTIVIDAD

La inclusión como condición especial será efectiva desde que se somete la certificación con las pruebas asociadas y el diagnóstico definitivo, según los criterios de inclusión arriba esbozados.

DURACIÓN DE LA CUBIERTA ESPECIAL

La cubierta especial durara mientras el paciente se mantenga suscrito al Plan de Salud del Gobierno de PR.

CUBIERTA ESPECIAL: Cubre.

1. Todos los servicios, pruebas y procedimientos médicamente necesarios de seguimiento por un oftalmólogo o dermatólogo para el manejo de la condición una vez establecido el diagnóstico.

2. En los casos de Síndrome de Hermansky-Pudlak y Chediak-Higashi, se cubrirán, además, los servicios, pruebas y procedimientos ofrecidos por un hematólogo.

3. Medicamentos prescritos por: oftalmólogos, dermatólogos y en los casos del Síndrome de Hermansky-Pudlak, aquellas prescritas por hematólogos, neumólogos para tratar condiciones o complicaciones en el manejo y prevención de complicaciones en esta población.

4. Lentes y espejuelos especialmente prescritos para protección, prevención y mejora de la visión, según los parámetros de cantidad y costo de estos establecidos por el Plan Vital.

   Se entiende que esto debe de incluir al menos espejuelos recetados cada dos años o cuando ocurran cambios significativos de visión, hasta un costó máximo por espejuelos que no excederá los $ 400.00 por unidad.

5. Cremas de protección solar específicas para prevención de complicaciones por exposición a los rayos ultravioletas. Estas lociones o cremas tienen que ofrecer un factor de protección solar SPF de 50 o más y proteger contra rayos ultravioletas A y B (UVA y UVB)

   La recomendación es de unas 24 onzas al mes (tres (3) botellas de 8 oz. /mes).

LIMITACIONES Y EXCLUSIONES

Tratamiento y manejo dentro de la Jurisdicción de Puerto Rico.

Terapias experimentales.

Tratamientos de modificación Genética.

Todas la limitaciones o exclusiones que apliquen al PSG

ACCESO A TRATAMIENTO

Según dispuesto en la cubierta especial, no se requerirán referidos ni contrafirma del médico primario para visitas y seguimientos con los especialistas: oftalmólogos y dermatólogos y, en los casos de Hermansky-Pudlak y Chediak-Higashi, las visitas al hematólogo.

Referencias:

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2. Hermansky and Pudlack: Albinism associated with Hemorrhagic Diathesis and unusual pigmented reticular cells in de bone marrow. Report of two cases with histochemical studies. Blood 1959; 14 (2): 162-169.
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4. Mayo Clinic: Albinism
5. Medline Plus. Ultima revisión 10/26/2017
6. NOAH - The National Organization for Albinism and Hypopigmentation PO Box 959, East Hampstead, NH 03826-0959
7. © Orphanet version 5.33.0 - Last updated: 2020-01-20
8. Power et al. Orphanet Journal of Rare Diseases (2019) 14:52
9. ¿Qué es el albinismo? Lluís Montoliu y Ana Yturralde (ALBA, 2018)
10. Senado de Puerto Rico. P. del S. 1127 18 de octubre de 2018

11. Senado de Puerto Rico. P. del S. 247 de 18 de marzo de 2021
12. Síndrome de Hermansky-Pudlak en Puerto Rico, características dermatológicas. Sánchez, Néstor, Santos Malavé Gabriel, Izquierdo, Natalio. Galenus, vol. 88/año 14/número 1 febrero-marzo 2021, págs. 32-34.
13. Treat Oculocutaneous Albinism with Gene Therapy Journal of Advances in Biology & Biotechnology 16(3): 1-12, 2017; Article no. JABB.38504 ISS: 2394-1081
14. Yang et al. BMC Medical Genetics (2019) 20:106; Genetic analyses of oculocutaneous albinism types 1 and 2 with four novel mutations

Mandated and Uniform Protocol for Conditions Included in Special Coverage

Initiation:

Any primary or specialist physician who have evaluated a patient may submit a request for Register subject to having available all required documentation for said condition. The insurer shall make a determination of approval or denial of registration and inform this decision in writing to the insured and the physician requesting the registration. If the physician requesting the registry is not the primary physician of the insured, the insurer shall send a copy of the determination to the primary care physician. The insurance company will make a final determination on the application for special coverage in a 72-hour period, after receiving the complete documentation as required by this Protocol for each condition.

Once a Provider supplies all the required information for the Contractor to process a registration and the Contractor processes such information, Special Coverage shall take effect retroactively as of the date the Provider reaches a diagnosis, including documentation of test results, for any condition included in Special Coverage. In case Information is submitted to the Contractor after the diagnosis was reached, coverage can be made retroactive up to sixty (60) Calendar Days before the date on which Provider submitted the registration request. (Contract Section 7.7.5)

Reactivation: Any insured who have lost eligibility for PSG for over one year period, will be required a new certification by the primary care physician that evidence current treatment plan to be reactivated in the special coverage. Any insured that loses its eligibility for a period less than 12 months, will be register without documents or additional certifications, unless there is any other limit for the specific condition.

Risk allocation*: the distribution of the special coverage between insurer and primary medical groups risk is defined in the following table. The same may be modify at the request of the insurance company subject to prior review and approval by ASES.

Notes:

1. **Covered medications are those included in the pharmacy benefit and ASES drug formulary (FMC).**
2. **The codes or diagnoses by themselves do not grant inclusion into a temporary special condition list. They must be in compliance with the criteria for inclusion as specified in the column named: Criteria for inclusion in the coverage**

Special Condition	Definitive diagnosis criteria for inclusion in the coverage	Special Coverage Effectiveness and Duration	Services included in Special Coverage	Risk Allocation*
1. Aplastic Anemia	1-Diagnosis certification by a hematologist/oncologist with treatment plan 2- Evidence of: a. Absolute Neutrophils Count b. Platelets Counts c. Reticulocytes Counts d. Results of bone Marrow aspiration or biopsy	**Effectiveness** = From the date of the diagnosis by the hematologist/oncologist or date the biopsy was performed if its reading establishes the definitive diagnosis. **Duration**= Special coverage will begin from the date the definitive diagnosis is established. Special cover will be in effect as long as the insured is eligible in the PSG	1. All hospital services, emergency room or medical specialist services provided with primary diagnosis of Aplastic Anemia. 2. All medical services provided or ordered by the hematologist/oncologist 3. Medication prescribed by the oncologist/ hematologist and specific to treat the condition.	**Insurer:** Medical services and medications as defined for the special coverage condition in this document. **GMP/PCP:** Will receive the monthly capitation corresponding to the insured.
2. Rheumatoid Arthritis	1-Diagnosis certification by the rheumatologist in accordance with the criteria established by the American College of Rheumatology. (The insurance company will provide a sheet with the criteria and treatment plan to be fill by the specialist.) . **American College of Rheumatology (ACR)/European Alliance of Associations for Rheumatology (EULAR) criteria**	**Effectiveness** = From the date of the diagnosis by the rheumatologist. **Duration** = Special cover will be in effect as long as the insured is eligible in the PSG	1. All hospital services, emergency room or medical specialist services provided with primary diagnosis of Rheumatoid Arthritis. 2. All medical services provided or ordered by the rheumatologist. 3. Medication prescribed by the rheumatologist and specific to treat the condition, including DMARD.	**Insurer:** Medical services and medications as defined for the special coverage condition in this document. **GMP/PCP** – Will receive the monthly capitation corresponding to the insured.

	1. Inflammatory arthritis affecting three or more joints. 2. Positive rheumatoid factor (RF) and/or anti-citrullinated peptide/protein (ACPA) functions, as evidence of anti-cyclic citrullinated peptide (anti-CCP) functions. 3. Elevated C-reactive protein or erythrocyte sedimentation rate. 4. Diseases with similar clinical features have been excluded, in particular psoriatic arthritis, acute viral polyarthritis, polyarticular gout or calcium pyrophosphate deposition disease, and systemic lupus erythematosus (SLE). 5. The duration of the symptoms is more than six weeks			
3. Autism **a. Provisional Coverage**	a. Certification of risk by the primary care physician and evidence of the screening tool utilized.	**Provisional Special Coverage:** a. **Effectiveness:** If the risk of developing the condition is confirm using the instruments established in the Protocol of Autism from the	**Provisional Special Coverage:** **a.** Diagnostic evaluation according to the Protocol of the Dept. of Health that includes family history, development and health,	a. **Insurer –** All services rendered by providers qualified for diagnostic evaluation.

	Codes to be used during the provisional coverage: 1. **R63.50** Unspecified lack of expected normal psychological development in childhood 2. **R62.0** Delayed Milestone in childhood 3. **F88** Other disorders of psychological development 4. **F80.2** Mixed receptive and expressive language disorders	Department of Health, the primary care physician will complete the registration form for provisional special coverage and send it to the insurer. Once the provisional special coverage for autism is activate, a referral or authorization from the primary care physician to access the services of a qualified provider for the diagnostic evaluation process will not be required. **Duration:** The provisional coverage will last for six months. If the evaluation process is not completed, the provisional coverage may be renew for six additional months. **b. Effectiveness:** From the date of the diagnosis certification by one of the listed professionals, the effective date will be the earliest certification date. **Duration:** Special coverage will be valid, provided the insured eligibility to the PSG, until 21 years of age. After 21 years, to continue in the special coverage, a certification by a neurologist or psychiatrist establishing the need for the condition management and treatment as an adult is required.	interview with tutors on the skills, behavior, communication and social interactions of the person, observation of the conduct of the person in interaction with others and own age play and socialization activities and the results of the most recent version of at least one instrument to document current behaviors. **b.** Medical services rendered or ordered by the psychiatrist, psychologist, neurologist, or any other qualified provider according to the Protocol of Autism from the Department of Health of PR will not require referral from the primary physician. Medicines for the specific management of the condition, prescribed by a qualified provider, will not require PCP authorization.	**GMP/PCP** – Will receive the monthly capitation corresponding to the insured. **b. Insurer:** Medical services and medications as defined for the special coverage condition in this document. **GMP/PCP** – Will receive the monthly capitation corresponding to the insured.
b. Permanent Special Coverage	**b.1.** Diagnosis certification by a clinical psychologist, school psychologist, counselor psychologist, neurologist, psychiatrist or a pediatrician development specialist. Professionals should have training or experience in the area of Autism, as required by the Protocol of Autism from the Department of Health of PR. **b. 2** Evidence of the relevant screening tests according to the Protocol of Autism from the Department of Health of PR.			

4. Cancer	1. Diagnostic certification with stage, by a hematologist/ oncologist or specialist physician in charge of the management of the condition, treatment plan with estimated start and completion dates. The insurer shall provide a specific form to be used as the Registry Application and Cancer Certification to be completed by the specialist. 2-Evidence of diagnosis by biopsy result. 3- In cases where the diagnosis cannot be confirmed by a pathology study, evidence of diagnostic studies of CT, MRI, PET Scan, ultrasonography supporting diagnosis or stage will be taken into consideration.	**Effectiveness** = from the date of certification of the diagnosis by the hematologist/oncologist or the biopsy date if its results establishes the definitive diagnosis. **Duration =** until the end of active treatment of the condition with radiotherapy or chemotherapy. All insured will receive a certification of registration until the date in which the insured meets their surgical treatment, chemotherapy and/or radiation therapy. The insured will have the benefit of covered visits to his oncologist/hematologist to a maximum of one year. At the end of the year, if needed, the hematologist/oncologist may perform a request for extension of registration documenting the condition stage and the treatment plan for next year. A temporary register up to a maximum of 30 days shall be granted to receive documentation on the Cancer Registration Extension form provided by the insurer. If this process is not completed, the insured will automatically lose its registration for special coverage. In cases of prostate cancer, treatment with hormonal chemotherapy will qualify the member to continue active in the cancer registry. Their visits to the urologist and medical orders and treatment ordered by this specialist (urologist) will be cover.	1. All hospital services, emergency room or medical specialist services provided with primary diagnosis of Cancer. 2-All medical services provided or ordered by the hematologist/oncologist. . 3- Medications prescribed by the hematologist/oncologist specific to treat the cancer condition.	**Insurer:** Medical services and medications as defined for the special coverage condition in this document. **GMP/PCP –** – Will receive the monthly capitation corresponding to the insured.

		In the cases of breast cancer, once active treatment with radiotherapy and chemotherapy ends, they will no longer remain in the registry. However, patients receiving treatment with anti-estrogens will continue being consider under cancer special coverage.		
5. Skin Cancer: Carcinoma IN SITU	- Positive Biopsy Report	**Effectiveness**: Special coverage in skin cancer and carcinoma in situ will only apply to the surgery day. **Duration:** the day or days for surgical removal and all services on said day and any other radiotherapy treatment used any time.	Surgical removal and all related services on said day and any other subsequent radiotherapy/chemotherapy treatment.	**Insurer:** Medical services and medications as defined for the special coverage condition in this document. **GMP/PCP –** – Will receive the monthly capitation corresponding to the insured.
6. Skin Cancer such as Invasive Melanoma or squamous cells with evidence of metastasis.	- Positive biopsy or pathology - Special studies like CT Scan, MRI, Sonogram - Registry certification completed by a dermatologist or a hematologist/oncologist.	**Effectiveness**: From the date the diagnosis is established. **Duration** = until the end of the active treatment of the condition with radiotherapy or chemotherapy. All insured will receive a certification of registration for up to a year. At the end of the year, if needed, the dermatologist or hematologist/oncologist may request an extension of registration documenting the condition stage and the treatment plan for next year. A temporary register up to a maximum of 30 days shall be granted to receive documentation on the Cancer Registration Extension form provided by the insurer. If this process is not	1. All hospital services, emergency room or medical specialist services provided with primary diagnosis of indicated Skin Cancer. 2-All medical services provided or ordered by the dermatologist or hematologist/oncologist. 3- Medications prescribed by the dermatologist or hematologist/oncologist specific to treat the cancer condition.	**Insurer:** Medical services and medications as defined for the special coverage condition in this document. **GMP/PCP:** Will receive the monthly capitation corresponding to the insured.

		completed, the insured will automatically lose its registration for special coverage.		
7. Chronic Renal Disease **Level 1 and 2** **Level 3 and 4** **Level 5**	The Glomerular Filtration Rate (GFR) is used. Evidence of recent results of Creatinine in blood and age, sex and race of the insured. **Level 1:** GFR over 90, ICD-10-N18.1 **Level 2:** GFR between 60 to 89, ICD-10-N18.2 **Level 3:** GFR between 30 to 59, ICD-10-N18.3 **Note: Starting on October 2020 the ICD-10 Codes for CKD3 will change. N18.0 will no longer be used. Subcategories of CKD3 will be identified as follows:** ***N18.30 Chronic kidney disease, stage 3 unspecified** ***N18.31 Chronic kidney disease, stage 3a** ***N18.32 Chronic kidney disease, stage 3b** **Level 4:** GFR between 15 to 29, ICD-10-N18.4	**Level 1 and 2:** Does not qualify for registry under special coverage. **Level 3 and 4:** Qualifies for special coverage registry. **Effectiveness**: From the date the diagnosis is established. **Duration =** As long as the insured is eligible in the PSG. **Level 5:**	**GMP/PCP**: Levels 1and 2 are total risk of GMP. **Level 3 and 4**-The insurer assumes the nephrologist visits (without referrals), renal laboratory and diagnostic studies ordered by this specialist, peripheral vascular studies to document hemodialysis access and drugs ordered by the nephrologist, related to the condition and limited to immunosuppressants, erythrocytes stimulants, Megace, renal antidotes and systemic corticosteroids **Level 5-**All services covered by the PSG as long as the insured is active in the Special Coverage Registry**.**	**GMP/PCP**: Levels 1and 2 are total risk of GMP. **Level 3 and 4:** **Insurer**: All medical services provided or ordered by nephrologist from the date of effectiveness of the coverage. Additionally including: - insertion of catheters for dialysis - surgeries for arteriovenous (AV) fistulas -Administration of hematopoietic agents - blood transfusions **GMP/PCP** **Level 3 and 4:** Will receive the monthly capitation corresponding to the insured. **Level 5**: **Insurer:** Once the registration for chronic kidney condition is

	Level 5: GFR less than 15 ICD-10-N18.5 ICD-10-N18.6 (ESRD)	**Effectiveness**: From the date the diagnosis is established. **Duration =** As long as the insured is eligible in the PSG		authorized, the insured received a notice by mail, indicating the changes in the coverage or the change of the GMP to one of the Renal-GMP (Dialysis Center). The change of GMP will be effective the month in which the change request is done. From this moment, the monthly capitation to the GMP for this insured is discontinued. The risk of the services received by the insured prior to the exchange of GMP or registration of the insured will be at the risk of the GMP, except those dealing directly with dialysis. Outpatient services, except emergency, provided to the insured in the Renal GMP have to be coordinated by the nephrologist, who will become the primary physician of the insured. **GMP/PCP:** **Level 5** – Will not receive monthly capitation for the insured.
8. Scleroderma	1. Diagnosis certification by the rheumatologist including signs and symptoms supporting the diagnosis. 2. Evidence of a positive ANA Test > or equal to1:80 dil	**Effectiveness**: From the diagnosis certification date by the rheumatologist. **Duration =** As long as the insured is eligible in the PSG	1. All hospital services, emergency room or medical specialist services provided with primary diagnosis of Scleroderma.	**Insurer:** Medical services and medications as defined for the special coverage condition in this document.

	3. Positive skin biopsy The insurer will develop a Registry form for this condition to be completed by the specialist certifying the condition, the criteria used to establish the diagnosis and the treatment plan.		2. All medical services provided or ordered by the rheumatologist. 3. Medication prescribed by the rheumatologist and specific to treat the condition.	**GMP/PCP:** Will receive the monthly capitation corresponding to the insured.
9. Multiple Sclerosis (MS) and Amiotrophic Lateral Sclerosis (ALS)	1. Certification of the diagnosis by a neurologist confirming condition and plan of treatment 2. Evidence of relevant diagnostic studies performed to reach diagnosis such as: MRIs, EMG, Evoked potentials, NCS, lumbar punction, Genetic studies, etc.	**Effectiveness**: From the date a definitive diagnosis is certified, and a treatment plan is established by the neurologist. **Duration =** As long as the insured is eligible in the PSG	1. All hospital services, emergency room or medical specialist services provided with primary diagnosis of MS or ALS. 2. All medical services provided or ordered by the neurologist. 3. Medication prescribed by the neurologist and specific to treat the condition.	**Insurer:** Medical services and medications as defined for the special coverage condition in this document. **GMP/PCP:** Will receive the monthly capitation corresponding to the insured.
10. Cystic Fibrosis	1. Sweat test 2. Evidence of treatments 3. Diagnosis certification by a pneumologist.	**Effectiveness**: From the date a definitive diagnosis is certified, and a treatment plan is established by the pneumologist. **Duration =** As long as the insured is eligible in the PSG	All services covered by the PSG as long as the insured is active in the Special Coverage Registry**.**	**Insurer**- All medically necessary services cover by the PSG. **GMP/PCP:** Monthly capitation does not apply for this insured.
11. Hemophilia	1. Certification of diagnosis by a hematologist 2. Evidence of relevant studies and test	**Effectiveness**: From the date a definitive diagnosis is certified, and a treatment plan is established by a hematologist. **Duration =** As long as the insured is eligible in the PSG	1- All hospital services, emergency room or medical specialist services provided with a diagnosis of hemophilia. 2-All medical services provided by the hematologist.	**Insurer:** Medical services and medications as defined for the special coverage condition in this document. **GMP/PCP:**

			3-Medications prescribed by the hematologist specifics to treat the condition and anti-hemophilic drugs administered to the insured.	Will receive the monthly capitation corresponding to the insured.
12. Leprosy	1. Evidence of skin biopsy result 2.Infection positive cultures 3. Diagnosis certification by an infectologist or a dermatologist.	**Effectiveness =** starts from the date of certification, which establishes the definitive diagnosis by the infectious disease specialist or a dermatologist. **Duration**= It ends when the treatment is complete.	1. All hospital services, emergency room or specialist, cultures, and biopsies of follow-up, provided with a diagnosis of leprosy. (ICD-10 A30) 2. All medical services provided by the infectious disease specialist or dermatologist. 3. Medications prescribed by the infectious disease specialist or dermatologist.	**Insurer:** Medical services and medications as defined for the special coverage condition in this document. **GMP/PCP:** Will receive the monthly capitation corresponding to the insured.
13. Systemic Lupus Erythematosus (SLE)	1-Diagnosis certification by a rheumatologist with evidence of the following tests: ANA Test, DS-DNA, Anti Sm y Anti Phospholipids.	**Effectiveness =** from the date of certification establishing the definitive diagnosis by the rheumatologist **Duration =** As long as the insured is eligible in the PSG	1. All hospital services, emergency room or medical specialist services provided with primary diagnosis of SLE. 2. All medical services provided or ordered by the rheumatologist. 3. Medication prescribed by the rheumatologist and specific to treat the condition of SLE.	**Insurer:** Medical services and medications as defined for the special coverage condition in this document. **GMP/PCP:** Will receive the monthly capitation corresponding to the insured.

14. Children with Special Health Needs	Complete the Registration Form for children with special health care needs by the primary care physician with evidence of the condition according to the list of diagnoses included by ASES as an attachment to the contract, entitled "Conditions to include patients in the Register of Children with Special Health Needs", revision of June 2015. Medical evidence will consist of relevant laboratories or tests, evidence of current treatment, diagnosis certifications by specialist physicians consulted and others.	**Effectiveness=** From the diagnosis certification date **Duration** = depends on whether the condition is temporary or permanent. The case manager will determine based on the Protocol established by the insurer the Registry duration, provided that the insured is under 21 years old.	As defined in the Conditions List revised on June 2015.	Refer to the listing of diagnosis codes of the conditions for Children with Special Needs Registry.
15. Obstetric	Obstretic Registry Form Certification of pregnancy by the obstretric gynecologist	**Effectiveness:** After registration, a certification of the special coverage will be mail to the insured. **Duration:** Registration will be effective since the estimated day of conception according to certification provided by the obstetrician and will continue to be effective until 56 days after the delivery date, provided this occur after the 20th week. If pregnancy ends in miscarriage before week 20, will only granted 30 days after the event.	All services covered by the PSG as long as the insured is active in the Special Coverage Registry**.** **Sterilization**: Sterilization carried out in a separate admission, after childbirth or caesarean section, will be responsibility of the primary medical group, therefore it will require referral from the PCP **Newborn:** newborn children will be cover as long as the mother have eligibility for the PSG, and until the Obstetrics Registration in in effect (56 days of the date of birth) at risk of the insurance company. Under the Obstetric Registry coverage, the assistance of the pediatrician during delivery by caesarean section or high risk and routine care for the	**Insurer:** All cover medical services and medications as long as the insured is active under this special coverage category. **GMP/PCP:** Will not receive monthly capitation for the insured. **Newborn:** per capita payment shall be paid for the newborn once the mother is out of the registration or the newborn is certified by the mother, whichever occurs first.

			newborn in the hospital (nursery room) are part of the obstetrics special coverage.	
16. Tuberculosis (Tb)	Pneumologist Certification with treatment plan and evidence of: 1- Tb test result 2- Chest radiology findings 3- Samples of sputum or bronchial wash for Acid-Fast Basillus (AFB) and culture for Mycobacterium tuberculosis. 4- Biopsies of the affected area, if applicable. 5- HIV test results	**Effectiveness =** from the date of certification establishing the definitive diagnosis by the pneumologist. **Duration:** Coverage will be variable, depending on the duration of the treatment, which can fluctuate between six (6) months to (1) year, depending on the plan of treatment certified by the pulmonologist. After the first year, if the patient requires continuing treatment, a re-evaluation of the case by the pulmonologist will be requested and according to the new plan of treatment, special coverage may be extended.	-Medical services related to the condition, follow-up, complications, complications of the diagnostic procedure and treatment shall be at the risk of the insurer from the date of effectiveness of the special coverage. -Special coverage includes medications to treat or control the special condition or conditions that may arise as part of diagnostic studies performed or from complications of the disease. -Chest radiology for follow up until the treatment is completed will be responsibility of the insurer. **Department of Health of PR covers:** - **Tuberculin** - **Culture** - **Bronchial washing** - **Medical treatment**	**Insurer:** Medical services and medications as defined for the special coverage condition in this document. **GMP/PCP:** Will receive the monthly capitation corresponding to the insured.

17. HIV/AIDS	Evidence of the result of any of the following laboratories; 1-Western Blot positive 2- positive HIV Viral load 3- positive 4th generation test with validation of the subtypes of antibody or Antigen for acute infection. The registration may be requested by one of the following providers: -Primary Care Physician -HIV/AIDS Clinics Physician -VIH/AIDS Clinics Case Manager	**Effectiveness =** from the date of certification establishing the definitive diagnosis **Duration =** As long as the insured is eligible in the PSG	1. All hospital services, emergency room or medical specialist services provided with primary diagnosis of HIV/AIDS. 2-All medical services provided or ordered by HIV/AID treaters. 3- Medications prescribed by the HIV/AID treaters specific to treat the HIV/AID condition.	**Insurer:** Medical services and medications as defined for the special coverage condition in this document. **GMP/PCP** – Will receive the monthly capitation corresponding to the insured.
18. Adults with phenylketonuria (PKU)	When the special coverage is a continuation to the coverage under children with special conditions, once the beneficiary reaches age 21, no additional evidence is required. The evidence that qualifies he/she as a child, serves the purpose for the continuation of coverage under the category of adult PKU. If it is not a continuation of coverage, the registry has to be request by the geneticist and shall include a treatment history and evidence of the result of the genetic study.	**Effectiveness:** it is a continuation of the registry under children with special conditions, after the beneficiary reaches age 21. **Duration =** As long as the insured is eligible in the PSG	1. All hospital services, emergency room or medical specialist services provided with primary diagnosis of PKU. 2. All medical services provided or ordered by the geneticist. 3. Medication prescribed by the geneticist and specific to treat the condition of PKU.	**Insurer:** Medical services and medications as defined for the special coverage condition in this document. **GMP/PCP:** Will receive the monthly capitation corresponding to the insured.

19. Pulmonary Hypertension	Diagnosis certification and treatment plan by the Pneumologist or Cardiologist and evidence of supporting test(s).	**Effectiveness =** from the date of certification establishing the definitive diagnosis by the pneumologist or cardiologist. **Duration =** As long as the insured is eligible in the PSG	1. All hospital services, emergency room or medical specialist services provided with primary diagnosis of Pulmonary Hypertension or its complications. 2. All medical services provided or ordered by the pneumologist or cardiologist to treat the condition or its complications. 3. Medication prescribed by pneumologist or cardiologist to treat the condition or its complications.	**Insurer:** Medical services and medications as defined for the special coverage condition in this document. **GMP/PCP:** Will receive the monthly capitation corresponding to the insured.
20. Post-Transplant **Nota:** EXCLUYE TRASPLANTE DE CÓRNEA, DE HUESO Y DE PIEL.	The primary care physician or the specialist (nephrologist, pneumologist, cardiologist, hepatologist or gastroenterologist) must submit: • A certification of the post transplant status including the diagnosis and transplant date • Treatment plan with starting dates • Specific immunosuppressors, doses and route of administration.	**Effectiveness** = from the date of certification and treatment plan **Duration**: Special cover will be in effect as long as the insured is eligible in de PSG	1. All hospital services, emergency room or medical specialist services provided related to the primary condition of post-transplant or its complications. 2. All medical services provided or ordered by the specialist or primary care physician to treat the post-transplant condition or its complications. 3. Medication prescribed by the specialists or primary care physician to treat the post-transplant condition or its complications.	**Insurer**- All medically necessary services cover by the PSG. **GMP/PCP:** Will receive the monthly capitation corresponding to the insured.
21. HCV (Chronic Hepatitis C) (Refer to **"Policy for the management of patients**	For its registry will be necessary to submit diagnosis certification including evidence of the following:	**Effectiveness=** From the date of registration with required certification and test results.	1. Direct access to the specialist or subspecialist that handles condition without referral of the PCP. 2. Treatment with the direct-acting antiviral drug (DDA) as established	**Insurer**- Medical services as defined for the special coverage condition in this document. Including but **not limited to**: Laboratories, (CMP,

diagnosed with Chronic Hepatitis-C under the GHIP" and to CN 20-0326)	• Positive result for HCV antibody (Ab) test and • Positive Quantitative RNA test • Treating physician should document and submit the treatment plan with estimated start and completion dates. • Treating physician should include in the registry, documents of letter of willingness to be treated from the beneficiary and agreements to start treatment immediately upon Registry in Special Condition Registry**.**	**Duration**= HCV special coverage will be in effect since the time the patient is registered on this special coverage until six (6) months <u>after</u> completing treatment with the direct-acting antiviral drug (DDA) with evidence of sustained virological response not detected. If after six (6) months after completion of treatment, there is no evidence of sustained virological response, then the Gastroenterologist or treating physician <u>MUST</u> document next step of management and treatment with specific start and completion dates. Otherwise the Beneficiary will revert to regular coverage and will be discontinued from special registry and coverage	under the Coverage of medication of ASES without countersignature of the PCP. 3. Medically Necessary Laboratories for the condition without referral of the PCP. 4. Imaging, sonography, MRI, CT or any other radiological imaging medically necessary for the condition without referral of the PCP.	PT & INR, CBC, Renal function test's, genotype, RNA quantitative, resistant test as needed, radiological imagines (sonogram, =with and w/o elastography, Liver CT and MRI if clinically indicated)and or any other medically necessary laboratories or tests to identify gradation and estimated degree of liver fibrosis in Hepatitis C, including liver biopsy with or w/o imaging guidance, & pathology report. Also included are the visits to Gastroenterologist or other specialized authorized physician as described in the **"Policy for the management of patients diagnosed with Chronic Hepatitis-C under the GHIP"** Laboratories, tests, imaging studies and interventional radiologist evaluation, biopsy and pathological report are covered from the moment the patient is included in the special coverage and until discharged from the special coverage inclusion. The recommended follow up during the medical treatment is included in the **"Policy for the management of patients diagnosed with Chronic**

				Hepatitis-C under the GHIP" as guidelines. (see pages 22-23). **GMP/PCP:** Will receive the monthly capitation corresponding to the insured. **ASES:** Pharmacological treatment with direct-acting antiviral drug (DDA).

22. Congestive Heart Failure (CHF): Class III and Class IV, NYHA. **ICD 10 Codes:** I50 Heart failure I50.1 Left ventricular failure, unspecified I50.2 Systolic (congestive) heart failure I50.20 Unspecified systolic (congestive) heart failure I50.22 Chronic systolic (congestive) heart failure I50.84 End stage heart failure	The treating cardiologist must fill a certificate stating the diagnosis of CHF with reduced Ejection Fraction (HFrEF) and document an Ejection Fraction (EF) equal or less than 30% and report with objective evidence findings and treatment offered to the beneficiary so far, until the date of referral. Must state that the Beneficiary is a real candidate for heart transplant and document at least **one (1)** of the followings: 1. Left Ventricular Ejection Fraction (LVEF) equal or less than 30%. 2. Recurrent or frequent hospitalizations because of decompensated Heart Failure. 3. Symptomatic CHF despite optimization of available medications and or the use of medical devices for treatment or compensation of CHF. (LVAD) or Left Ventricular Assist Devices. 4. Continued and prolonged large doses of, or frequent increase in, dosages of diuretic medications. 5. Dependant on positive inotropics medications. **AND:** • Absence of severe right ventricular dysfunction and tricuspid regurgitation.	**Effective date of inclusion:** **Special Temporary Coverage as special condition** will be effective when all the documentation in the second column is submitted by the treating cardiologist and is preliminary evaluated **and** accepted by the Transplant Center for further evaluation as a potential or possible candidate for heart transplant**.** **Duration of Coverage:** This Special Temporary Coverage will last only for a MAXIMUM non- extendable period of four (4) months, commencing on the effective inclusion date, and will last for four months or until the Beneficiary is accepted for transplant or declined as a candidate for transplant whichever occurs first. After this timeframe, the beneficiary will return to the Regular Coverage without any further appeal.	**The following tests, laboratory tests or work up will be covered only ONCE during the Special Temporary Coverage Period:** -ABO type and Screen -CBC + differential -Glycosylated Hgb, -Lymphocyte Sub- Population Determination -CMP, -TSH, T3, T4, -Uric Acid blood levels. -Fasting Lipid Profile -Urinalysis, Urine Culture -Blood and Throat culture X1. -Urine Collection X 24 hrs. for creatinine clearance and total proteins -CMV -Toxoplasma -Varicella -Herpes Simplex -Measles -Rubella -Epstein Bar lgG & lgM -HIV -Hepatitis profile -RPR -Legionella Antibodies -Panel Reactive Antibodies -HLA A, B, DQ, DR -Nicotine in urine -Stool for OVA and Parasites -Stool for Occult Blood in patients 50 years old or older. -Pregnancy Test in female in reproductive age. -PSA (males > 40 años)	**MCO:** At risk of all studies, laboratories, and medical and other included evaluations according to the list in the left column during the period of four (4) months as described in column three. **GMP/PCP:** Will receive his monthly capitation during the special temporary coverage period. All studies, laboratory and medical evaluations will be given back to the Beneficiary and be available in electronic format to the treating cardiologist and PCP. All these evaluations will count toward quality requirements for PCP evaluations and CMO incentives as contracted with PCP.

			-Hair Toxicology tests (Patients with history of ilicit drug use) -MRSA Test -BNP Levels. **The following evaluation will be cover: To minimize duplication of services and studies, the evaluations will be done after all the laboratory results pertinent to the specialist who will evaluate the Beneficiary are available** • Neumologist • Neprologist • Infection disease • Dentist • Gynecologist • Urologist • Psychiatrist • Nutritional Evaluation.	

23. Primary Ciliary Dyskinesia (PCD) Or **Discinesia Ciliar Primaria (DCP)** Or **Síndrome de cilios inmóviles** Or **Immotile Ciliary Syndrome** Or **Síndrome de Kartagener** **ICD 10 Codes:** Q34.8 Q34.9 Q89.3	Referral for inclusión by: Pediatric age: - Pediatric pneumologist - Pediatric - Genetist - Immunologist Adult: - Pneumologist - Primary care physician (PCP0) **PLUS**, one of the following: a. Biopsy of ciliated tissue (usually from the nose or trachea) with analysis of ciliary ultrastructure. Or b. Genetic test showing two mutations known to cause PCD—one from each parent	**Effectiveness** = From the date of the diagnosis by one of the specialists listed in the left column or date the biopsy was performed and reported as positive for PCD **Duration** = Special cover will be in effect as long as the insured is eligible in the PSG	1. All hospital services, emergency room or medical specialist services provided with primary diagnosis of PCD 2. All medical services provided or ordered by the Neumologist, pediatric or adult, included all referral for evaluation with specialist and subspecialist for conditions related to the primary diagnosis of PCD or its complications. 3. Medication prescribed by the Neumologist and specific to treat the condition or its complications or mediations prescribed by one of the specialist or subspecialists treating or evaluating patients with primary diagnosis of PCD and or its complications.	**Insurer**: Medical services and medications as defined for the special coverage condition in this document and described in the clinical protocol for PCD **GMP/PCP**: Will receive the monthly capitation corresponding to the insured.

24. Cleft palate and or Cleft lips	a. Usually identified at birth, with physical examination, or prenatal with ultrasound. No need for special test for diagnosis. May be certified by obstetrician or neonatologist/ perinatologist and or pediatrician.	**Effectiveness**: Since diagnosis usually at birth or prenatal. **Duration**: Until condition is surgically repaired and or age 12. Then and only in those persons requiring further surgeries o management, will be re-certified for 5 years up to two times by ENT, Plastic surgeon or maxillofacial surgeon, with a step-by-step plan for pending surgeries.	All visits, evaluations, surgeries, therapies, and rehabilitation. 1. Surgeons who specialize in cleft repair, such as plastic surgeons and 2. Otolaryngologist 3. Oral surgeons 4. Pediatricians 5. Dentists pediatric 6. Orthodontists 7. Nurses 8. Audiologists 9. Speech therapists. 10. Geneticist and/or genetic counselors 11. Social workers 12. Psychologists Laboratories, radiological and nuclear studies Materials and equipment including headphones and/or audiological. Equipment. Required audiological studies Genetic studies Videonasopharyngoscopy nasometry Videofluoroscopy Bone implant or inserts **In the special condition protocol, the coverage of evaluations, services, laboratories and other radiological tests and others, will be limited to those that are covered by the Health Plan of the Government of Puerto Rico.**	**Insurer: All risk to insurer** **GMP/PCP: No risk and continues to receive PPMM.**

			Under no circumstances is it intended to include services, tests, laboratories, or evaluations that are not in the current coverage of the Health Plan of the Government of Puerto Rico.	

25. IBD (Inflammatory Bowel Diseases) **~ Enfermedad de Crohn** **~ Colitis ulcerativa** **~ Colitis microscópica** **<u>Crohn's disease : ICD-10</u>** **K50.xxx** **K50.0 small bowel** **K50.1 colon** **K50.8 small and large bowel** **K50.9 unspecified** **<u>Ulcerative colitis: ICD-10</u>** **K51.xxx** **K51.0 pancolitis** **K51.2 proctitis** **K51.3 rectosigmoiditis** **K51.5 left sided colitis** **K51.9 unspecified** **<u>Indeterminate colitis k52.3</u>**	1. A diagnostic certification by a gastroenterologist will be required **<u>AND</u>** 2. Endoscopy studies (colonoscopy / sigmoidoscopy **<u>AND</u>** 3. Reliable diagnostic tests (biopsy) and/or imaging studies that document it. **<u>AND</u>** 4. Evidence ruling out acute infectious etiology.	**Effectiveness**: The inclusion as a special condition will be effective from the time the certification is submitted with the associated tests and the definitive diagnosis, according to the inclusion criteria outlined above. **Duration**: The special coverage will last as long as the patient remains subscribed to the PR Government Health Plan.	1. All hospital, emergency or specialist medical services provided for management once the IBD condition has been diagnosed. 2. All medical services rendered or ordered by the gastroenterologist or rendered by another specialist referred by the gastroenterologist to evaluate or diagnose and/or treat related conditions or complications of IBD. Special mention is made without being exclusive of: Ophthalmology, surgery, dermatology, rheumatology, infectiology, radiological and imaging services, pathology, nutrition and dietetics, psychiatry, and psychology. 3. Including all surgeries related to the management, treatment, and complications of IBD. 4. Medications prescribed by gastroenterologist and specific to treat the condition and its symptoms and complications, including topical therapies, immunomodulators,	Insurer: Medical services and medications as defined for the special coverage condition in this document. GMP/PCP – will receive the monthly capitation corresponding to the insured.

			biologics, small molecules, and antibiotics, validated by evidence and medical practice. 5. Laboratory tests. **6. All supply and materials for ostomized patients secondary to complication or required appropriate management of IBD, including the following codes:** **A4361 OST FACEPLATE** **A4362 SKIN BARRIER; SOLID 4 FOUR OR EQUIVALENT; EACH** **A4364 ADHESIVE LIQUID OR EQUAL, ANY TYPE PER OZ.** **A4367 OST BELT EACH** **A4369 OST SKIN BARRIER LIQUID PER OZ** **A4371 OST SKIN BARRIER POWDER PER OZ** **A4385 OST SKN BARRIER SOLID 4X4 EXT W/O CONVXITY EA** **A4395 OST DEODORANT TO USE IN OSTOMY POUCH SOLID PER TABLET** **A4405 OST SKIN BARRIER, NON PECTIN BASED, PASTE PER OZ.** **A4406 OST SKIN BARRIER PECTIN-BASED PASTE PER OUNCE** **A4407 OST SKN BARRIER W/BUILT-IN CONVXITY 4X4 IN/< EA**	

			A4409 OST SKN BARR EXT W/O BUILT-IN CONVXTY 4X4 IN/4X4 IN EA **A4419 OST POUCH CLOS; BARRIER W/NON-LOCK FLNGE W/FLTR** **A4421 OSTOMY SUPPLY MISCELANEOUS** **A4422 OST ABSORBENT MATERIAL (SHEET/PAD/CRYSTAL PACKET) FOR USE IN OSTOMY POUCH TO THICKEN LIQUID STOMAL OUTPUT, EACH.** **A4450 TAPE NON-WATERPROOF PER 18 SQUARE INCHES** **A4452 TAPE WATERPROOF PER 18 SQUARE INCHES** **A4456 ADHESIVE REMOVER WIPES ANY TYPE EACHA** **A5120 SKIN BARRIER WIPES OR SWABS EACH** **THE PREVIOUS LIST IS NOT EXHAUSTIVE AS IT MAY INCLUDE SOME NOT LISTED ABOVE MEDICAL NEEDED SUPPLY.** **QUANTITY OF EACH SUPPLY IS ACCORDING TO MCS RECOMMENDED FOR EACH OSTOMY MATERIAL OR SUPPLY.**	

26. Oculocutaneous albinism, Hermansky-Pudlak syndrome (HSP), and Chédiak–Higashi syndrome (CHS). **ICD 10 Codes:** **E70.329** **Oculocutaneous albinism** **E70.331** **Hermansky-Pudlak síndrome** **E70.330** **Chediak-Higashi syndrome**	A diagnostic certification by one of the following specialists will be required: Dermatologist Ophthalmologist geneticist and Results of examinations or tests that support the diagnosis such as: -Genetic testing -Skin pigments -Ophthalmological studies In the case of syndromic OCA, it must also be accompanied by a certification by a hematologist.	**Effectiveness:** Inclusion as a special condition will be effective from the moment the certification is submitted with the associated tests and the definitive diagnosis, according to the inclusion criteria outlined above. **Duration:** The special coverage will last as long as the patient remains subscribed to the PR Government Health Plan.	1. All medically necessary follow-up services, tests, and procedures by an ophthalmologist or dermatologist for the management of the condition once the diagnosis has been established. 2. In the cases of Hermansky-Pudlak and Chediak-Higashi Syndrome, the services, tests and procedures offered by a hematologist will also be covered. 3. Medications prescribed by: ophthalmologists, dermatologists and in the cases of Hermansky-Pudlak Syndrome, those prescribed by hematologists, pulmonologists to treat conditions or complications in the management and prevention of complications in this population. 4. Lenses and spectacles specially prescribed for protection, prevention and improvement of vision, according to the quantity and cost parameters of these established by the Vital Plan. It is understood that this must include at least prescription glasses every two years or when significant vision changes occur, up to a maximum cost per eyeglass that will not exceed $400.00 per unit. 5. Specific sun protection creams to	Insurer: Medical services and medications as defined for the special condition, as well as diagnostic tests, laboratories and other studies required and ordered by the specialist dermatologist, ophthalmologist, geneticist and/or hematologist GMP/PCP: Will receive the monthly capitation corresponding to the insured.

			prevent complications from exposure to ultraviolet rays. These lotions or creams must offer an SPF sun protection factor of 50 or more and protect against ultraviolet A and B (UVA and UVB) rays. The recommendation is about 24 ounces per month (three (3) 8 oz. bottles/month).	