

Hemophilia Products – Factor VIII/VWF Complex: Alphanate, Humate-P®, Wilate (Intravenous)

Document Number: IC-0345

Last Review Date: 06/01/2023

Date of Origin: 12/16/2014

Dates Reviewed: 12/2014, 04/2015, 05/2015, 09/2015, 12/2015, 03/2016, 06/2016, 12/2016, 06/2017, 09/2017, 11/2017, 11/2018, 03/2019, 11/2019, 02/2020, 06/2021, 06/2022, 06/2023

I. Length of Authorization

Coverage is provided for 3 months and may be renewed thereafter, unless otherwise specified*.

Note: The cumulative amount of medication the patient has on-hand will be taken into account for authorizations. Up to 5 'on-hand' doses for the treatment of acute bleeding episodes will be permitted at the time of the authorization request.

**Initial and renewal authorization periods may vary by specific covered indication*

II. Dosing Limits

A. Quantity Limit (max daily dose) [NDC Unit]:

- N/A

B. Max Units (per dose and over time) [HCPCS Unit]:

- Alphanate: 55,200 billable units per 28 day supply
- Humate-P: 55,200 billable units per 28 day supply
- Wilate: 55,200 billable units per 28 day supply

III. Initial Approval Criteria ^{1-5,10}

Hemophilia Management Program
Requirements for half-life study and inhibitor tests are a part of the hemophilia management program. This information is not meant to replace clinical decision making when initiating or modifying medication therapy and should only be used as a guide.

Coverage is provided in the following conditions:

A. Alphanate, Humate-P ONLY

Hemophilia A (congenital factor VIII deficiency) †

- Diagnosis of congenital factor VIII deficiency has been confirmed by blood coagulation testing: **AND**
- Used as treatment in one of the following:

- Treatment and control of acute bleeding episodes (episodic treatment of acute hemorrhage); **OR**
- Perioperative management (**Authorization is valid for 1 month*); **OR**
- Routine prophylaxis to prevent or reduce the frequency of bleeding episodes; **AND**
 - Patient must have severe hemophilia A (factor VIII level of <1%); **OR**
 - Patient has at least two documented episodes of spontaneous bleeding into joints

Hemophilia Management Program
<ul style="list-style-type: none"> • If the request is for routine prophylaxis and the requested dose exceeds dosing limits under part II, a half-life study should be performed to determine the appropriate dose and dosing interval. • For members with a BMI ≥ 30, a half-life study should be performed to determine the appropriate dose and dosing interval. • For minimally treated patients (< 50 exposure days to factor products) previously receiving a different factor product, inhibitor testing is required at baseline, then at every comprehensive care visit (yearly for the mild and moderate patients, semi-annually for the severe patients)

von Willebrand disease (vWD) † Φ

- Diagnosis of von Willebrand disease has been confirmed by blood coagulation and von Willebrand factor testing; **AND**
- Used as treatment in one of the following:
 - Spontaneous and trauma-induced bleeding episodes; **OR**
 - Surgical bleeding prophylaxis during major or minor procedures in patients with vWD in whom desmopressin is either ineffective or contraindicated (**Authorization valid for 1 month*); **AND**
- Alphanate is not indicated for patients with severe (type 3) vWD undergoing major surgery **OR** treatment of spontaneous/trauma-induced bleeding episodes

Hemophilia Management Program
For minimally treated patients (< 50 exposure days to factor products) previously receiving a different factor product, inhibitor testing is required at baseline, then at every comprehensive care visit (yearly for the mild and moderate patients, semi-annually for the severe patients)

B. Wilate

Hemophilia A (congenital factor VIII deficiency) †

- Diagnosis of congenital factor VIII deficiency has been confirmed by blood coagulation testing; **AND**
- Used as treatment in one of the following:
 - On-demand treatment and control of bleeding episodes; **OR**

- Routine prophylaxis to prevent or reduce the frequency of bleeding episodes; **AND**
 - Patient must have severe hemophilia A (factor VIII level of <1%); **OR**
 - Patient has at least two documented episodes of spontaneous bleeding into joints

von Willebrand disease (vWD) † Φ

- Diagnosis of von Willebrand disease has been confirmed by blood coagulation and von Willebrand factor testing; **AND**
- Used as treatment in one of the following;
 - Perioperative management of bleeding (**Authorization valid for 1 month*); **OR**
 - On-demand treatment and control of bleeding episodes in at least one of the following:
 - Patients with severe vWD; **OR**
 - Patients with mild or moderate vWD in whom the use of desmopressin is known or suspected to be ineffective or contraindicated

Hemophilia Management Program
For minimally treated patients (< 50 exposure days to factor products) previously receiving a different factor product, inhibitor testing is required at, then at every comprehensive care visit (yearly for the mild and moderate patients, semi-annually for the severe patients)

† FDA Approved Indication(s); ‡ Compendia Recommended Indication(s); Φ Orphan Drug

IV. Dispensing Requirements for Rendering Providers (Hemophilia Management Program)

- Prescriptions cannot be filled without an expressed need from the patient, caregiver or prescribing practitioner. Auto-filling is not allowed.
- Monthly, rendering provider must submit for authorization of dispensing quantity before delivering factor product. Information submitted must include:
 - Original prescription information, requested amount to be dispensed, vial sizes available to be ordered from the manufacturer, and patient clinical history (including patient product inventory and bleed history)
 - Factor dose should not exceed +1% of the prescribed dose and a maximum of three vials may be dispensed per dose. If unable to provide factor dosing within the required threshold, below the required threshold, the lowest possible dose able to be achieved above +1% should be dispensed. Prescribed dose should not be increased to meet assay management requirements.
- The cumulative amount of medication(s) the patient has on-hand should be taken into account when dispensing factor product. Patients should not have more than 5 extra doses on-hand for the treatment of acute bleeding episodes.

- Dispensing requirements for renderings providers are a part of the hemophilia management program. This information is not meant to replace clinical decision making when initiating or modifying medication therapy and should only be used as a guide.

V. Renewal Criteria ^{1-3,4,5,10}

Coverage can be renewed based upon the following criteria:

- Patient continues to meet indication-specific relevant criteria identified in section III; **AND**
- Absence of unacceptable toxicity from the drug. Examples of unacceptable toxicity include: anaphylaxis and hypersensitivity reactions (e.g., angioedema, urticaria, tachycardia, chest tightness, hypotension, rash, nausea, vomiting, paresthesia, restlessness, wheezing, dyspnea, etc.), thromboembolic events (thromboembolism, pulmonary embolism), development of neutralizing antibodies (inhibitors), etc.; **AND**
- Any increases in dose must be supported by an acceptable clinical rationale (i.e., weight gain, half-life study results, increase in breakthrough bleeding when patient is fully adherent to therapy, etc.); **AND**
- The cumulative amount of medication(s) the patient has on-hand will be taken into account when authorizing. The authorization will allow up to 5 doses on-hand for the treatment of acute bleeding episodes as needed for the duration of the authorization; **AND**

Treatment and control of acute bleeding episodes/Treatment of spontaneous and trauma-induced bleeding episodes/On-demand treatment of bleeding episodes

- Renewals will be approved for a 6 month authorization period

Perioperative management of surgical bleeding/Surgical bleeding prophylaxis

- Coverage may NOT be renewed

Routine prophylaxis to prevent or reduce the frequency of bleeding episodes

- Renewals will be approved for a 12 month authorization period; **AND**
- Patient has demonstrated a beneficial response to therapy (i.e., the frequency of bleeding episodes has decreased from pre-treatment baseline)

VI. Dosage/Administration ¹⁻⁵

Alphanate

Indication	Dose
Control and prevention of bleeding Congenital Hemophilia A	The expected in vivo peak increase in FVIII level expressed as IU/dL (or % normal) can be estimated using the following formulas: - $Dosage \text{ (international units)} = body \text{ weight (kg)} \times desired \text{ FVIII rise (IU/dL or \% normal)} \times 0.5 \text{ (IU/kg per IU/dL)}$ OR

Indication	Dose
	<p>– IU/dL (or % of normal) = $[Total\ Dose\ (IU)/body\ weight\ (kg)] \times 2$</p> <p><u>Minor</u> FVIII:C levels should be brought to 30% of normal (15 IU FVIII/kg twice daily) until hemorrhage stops and healing has been achieved (1-2 days).</p> <p><u>Moderate</u> FVIII:C levels should be brought to 50% (25 IU FVIII/kg twice daily) until healing has been achieved (2-7 days, on average).</p> <p><u>Major</u> FVIII:C levels should be brought to 80-100% for at least 3-5 days (40-50 IU FVIII/kg twice daily). Following this treatment period, FVIII levels should be maintained at 50% (25 IU FVIII/kg twice daily) until healing has been achieved. Major hemorrhages may require treatment for up to 10 days. Intracranial hemorrhages may require prophylaxis therapy for up to 6 months.</p>
<p>Perioperative management Congenital Hemophilia A</p>	<p>Prior to surgery, the levels of FVIII:C should be brought to 80-100% of normal (40-50 IU FVIII/kg). For the next 7-10 days after surgery, or until healing has been achieved, the patient should be maintained at 60-100% of normal (30-50 IU FVIII/kg twice daily).</p>
<p>Control and prevention of bleeding and perioperative management von Willebrand Disease (VWD)</p>	<p>The ratio of VWF:RCo to FVIII in Alphanate varies by lot, so with each new lot, check the IU VWF:RCo/Vial to ensure accurate dosing.</p> <p><u>Minor</u> <u>Pre-operative/pre-procedure dose (Target FVIII:C Activity – 40-50 IU/dL):</u> Adults: 60 IU VWF:RCo/kg body weight. Pediatrics: 75 IU VWF:RCo/kg body weight. <u>Maintenance dose (Target FVIII:C Activity – 40-50 IU/dL):</u> Adults: 40-60 IU VWF:RCo/kg body weight at 8 to 12 hour intervals as clinically needed for 1-3 days. Pediatrics: 50-75 IU VWF:RCo/kg body weight at 8 to 12 hour intervals as clinically needed for 1-3 days.</p> <p><u>Major</u> <u>Pre-operative/pre-procedure dose (Target FVIII:C Activity – 100 IU/dL):</u> Adults: 60 IU VWF:RCo/kg body weight. Pediatrics: 75 IU VWF:RCo/kg body weight. <u>Maintenance dose (Target FVIII:C Activity – 100 IU/dL):</u> Adults: 40-60 IU VWF:RCo/kg body weight at 8 to 12 hour intervals as clinically needed for at least 3-7 days. Pediatrics: 50-75 IU VWF:RCo/kg body weight at 8 to 12 hour intervals as clinically needed for at least 3-7 days.</p>

Humate-P

Indication	Dose
Control and prevention of bleeding Congenital Hemophilia A	<p>One International Unit (IU) of Factor VIII (FVIII) activity per kg body weight will increase the circulating FVIII level by approximately 2.0 International Units (IU)/dL.</p> <p><u>Minor</u></p> <ul style="list-style-type: none"> – <u>Loading Dose:</u> Administer 15 IU FVIII:C/kg intravenously to achieve a FVIII:C plasma level of approximately 30% of normal; one infusion may be sufficient. If needed, half of the loading dose may be given once or twice daily for 1-2 days. <p><u>Moderate</u></p> <ul style="list-style-type: none"> – <u>Loading Dose:</u> Administer 25 IU FVIII:C/kg intravenously to achieve a FVIII:C plasma level of approximately 50% of normal, followed by 15 IU FVIII:C/kg every 8-12 hours for the first 1-2 days to maintain the FVIII:C plasma level at 30% of normal. Continue the same dose once or twice daily for up to 7 days or until adequate wound healing is achieved. <p><u>Major</u></p> <ul style="list-style-type: none"> – Initially administer 40-50 IU FVIII:C/kg intravenously, followed by 20-25 IU FVIII:C/kg every 8 hours to maintain the FVIII:C plasma level at 80-100% of normal for 7 days. Continue the same dose once or twice daily for another 7 days to maintain the FVIII:C level at 30-50% of normal.
Control and prevention of bleeding von Willebrand Disease (VWD)	<p>Administer 40-80 IU VWF:RCo intravenously (corresponding to 17 to 33 IU FVIII in Humate-P) per kg body weight every 8-12 hours. Adjust the dosage based on the extent and location of bleeding. Administer repeat doses as long as needed based on monitoring of appropriate clinical and laboratory measures.</p>
Perioperative management von Willebrand Disease (VWD)	<p><u>Loading Doses (to be administered 1 to 2 hours before surgery)</u></p> <p><u>Major</u></p> <ul style="list-style-type: none"> – <u>VWF:RCo Target Peak Plasma Level:</u> 100 IU/dL – <u>FVIII:C Target Peak Plasma Level:</u> 80-100 IU/dL <p>Calculation of Loading Dose:</p> <ul style="list-style-type: none"> – $(\text{Target peak plasma VWF:RCo level} - \text{baseline plasma VWF:RCo level}) \times \text{Body wt (kg)} / \text{IVR (in vivo recovery)}$ – If the IVR is not available, assume an IVR of 2.0 IU/dL per IU/kg and calculate the loading dose as follows: $(100 - \text{baseline plasma VWF:RCo}) \times \text{Body Weight (kg)} / 2.0$ <p><u>Minor</u></p> <ul style="list-style-type: none"> – <u>VWF:RCo Target Peak Plasma Level:</u> 50-60 IU/dL – <u>FVIII:C Target Peak Plasma Level:</u> 40-50 IU/dL <p>Calculation of Loading Dose:</p> <ul style="list-style-type: none"> – $(\text{Target peak plasma VWF:RCo level} - \text{baseline plasma VWF:RCo level}) \times \text{Body weight (kg)} / \text{IVR (in vivo recovery)}$

Indication	Dose
	<p>Emergency</p> <ul style="list-style-type: none"> - <u>VWF:RCo Target Peak Plasma Level: 100 IU/dL</u> - <u>FVIII:C Target Peak Plasma Level: 80-100 IU/dL</u> <p>Administer a dose of 50-60 IU VWF:RCo/kg body weight.</p> <p>Maintenance Doses</p> <p>The initial maintenance dose of Humate-P for the prevention of excessive bleeding during and after surgery should be half of the loading dose, irrespective of additional dosing required to meet FVIII:C targets. Subsequent maintenance doses should be based on the patient's VWF:RCo and FVIII levels.</p>

Wilate

Indication	Dose
Control of bleeding episodes von Willebrand Disease (VWD)	<p>Calculation of the required dose of VWF:RCo is based on the empirical finding that 1 IU VWF:RCo per kg body weight raises the plasma VWF activity by approximately 2% of normal activity or 2 IU/dL, using the following formula:</p> <ul style="list-style-type: none"> - $Required\ IU = body\ weight\ (kg) \times desired\ VWF:RCo\ rise\ (\%) (IU/dL) \times 0.5 (IU/kg\ per\ IU/dL)$ - $Expected\ VWF:RCo\ rise\ (\% \ of\ normal) = 2 \times administered\ IU / body\ weight\ (kg)$ <p>Adjust the dosage and frequency of administration to the clinical effectiveness in the individual patient.</p> <p>The ratio between VWF:RCo and FVIII activities in Wilate is approximately 1:1. The dosage should be adjusted according to the extent and location of the bleeding.</p> <p>Minor</p> <ul style="list-style-type: none"> - <u>Loading Dose:</u> Administer 20-40 IU/kg intravenously - <u>Maintenance Dose:</u> Administer 20-30 IU/kg intravenously every 12-24 hours, up to 3 days. VWF:RCo and FVIII activity trough levels > 30%. <p>Major</p> <ul style="list-style-type: none"> - <u>Loading Dose:</u> Administer 40-60 IU/kg intravenously - <u>Maintenance Dose:</u> Administer 20-40 IU/kg intravenously every 12-24 hours up to 5-7 days. VWF:RCo and FVIII activity trough levels > 50%.
Perioperative management of bleeding von Willebrand Disease (VWD)	<p>Calculation of the required dose of VWF:RCo is based on the empirical finding that 1 IU VWF:RCo per kg body weight raises the plasma VWF activity by approximately 2% of normal activity or 2 IU/dL, using the following formula:</p> <ul style="list-style-type: none"> - $Required\ IU = body\ weight\ (kg) \times desired\ VWF:RCo\ rise\ (\%) (IU/dL) \times 0.5 (IU/kg\ per\ IU/dL)$ - $Expected\ VWF:RCo\ rise\ (\% \ of\ normal) = 2 \times administered\ IU / body\ weight\ (kg)$ <p>Adjust the dosage and frequency of administration to the clinical effectiveness in the individual patient.</p> <p>Minor</p> <ul style="list-style-type: none"> - <u>Loading Dose:</u> Administer 30-60 IU/kg intravenously

Indication	Dose																										
	<ul style="list-style-type: none"> – <u>Maintenance Dose:</u> Administer 15-30 IU/kg intravenously or half of the loading dose every 12-24 hours until wound healing achieved, up to 3 days. VWF:RCo trough levels > 30% and peak levels 50%. <p>Major</p> <ul style="list-style-type: none"> – <u>Loading Dose:</u> Administer 40-60 IU/kg intravenously – <u>Maintenance Dose:</u> Administer 20-40 IU/kg intravenously or half the loading dose every 12-24 hours (at least 2 doses within the first 24 hours after the start of surgery) until wound healing achieved, up to 6 days or more. VWF:RCo trough levels > 50% and peak levels 100%. 																										
Control and prevention of bleeding/Routine Prophylaxis Congenital Hemophilia A	<p>Calculation of the required dose of Factor VIII is based on the empirical finding that 1 IU Factor VIII per kg body weight raises the plasma Factor VIII activity by approximately 2% of normal activity or 2 IU/dL when assessed using the one stage clotting assay. Use the following formula to determine the required dose:</p> <ul style="list-style-type: none"> – <i>Required IU = body weight (kg) x desired Factor VIII rise (%) (IU/dL) x 0.5 (IU/kg per IU/dL)</i> – <i>Expected Factor VIII rise (% of normal) = 2 x administered IU / body weight (kg)</i> <p>Dose and duration of therapy depend on the patient's weight, type, and severity of hemorrhage, FVIII level, and presence of inhibitors. Titrate dose and frequency to the patient's clinical response, individual needs, severity of deficiency, severity of hemorrhage, desired FVIII level, and presence of inhibitor, and the patient's clinical condition. Patients may vary in their pharmacokinetic (e.g., half-life, in vivo recovery) and clinical responses to Wilate.</p> <p>Routine Prophylaxis</p> <p>A guide for dosing as routine prophylaxis to reduce the frequency of bleeding is provided below. Exact dosing should be defined by the patient's clinical status and response.</p> <table border="1"> <thead> <tr> <th>Patients</th> <th>Recommended Dose (IU/kg)</th> <th>Frequency</th> </tr> </thead> <tbody> <tr> <td>Adolescents and adults</td> <td>20-40 IU/kg</td> <td>Every 2-3 days</td> </tr> </tbody> </table> <p>Treatment of Hemorrhages</p> <p>A guide for dosing in the treatment of hemorrhages is provided below. Exact dosing should be defined by the patient's clinical status and response.</p> <table border="1"> <thead> <tr> <th>Hemorrhage Type</th> <th>Recommended Dose (IU/kg)</th> <th>Frequency</th> <th>Duration of Therapy</th> </tr> </thead> <tbody> <tr> <td>Minor</td> <td>30-40</td> <td>Repeat every 12-24 hours</td> <td>At least 1 day, until bleed stops</td> </tr> <tr> <td>Moderate</td> <td>30-40</td> <td>Repeat every 12-24 hours</td> <td>3+ days, until bleed stops</td> </tr> <tr> <td>Major</td> <td>35-50</td> <td>Repeat every 12-24 hours</td> <td>3+ days, until bleed stops</td> </tr> <tr> <td>Life-Threatening</td> <td>35-50</td> <td>Repeat every 8-24 hours</td> <td>Until threat has resolved</td> </tr> </tbody> </table>	Patients	Recommended Dose (IU/kg)	Frequency	Adolescents and adults	20-40 IU/kg	Every 2-3 days	Hemorrhage Type	Recommended Dose (IU/kg)	Frequency	Duration of Therapy	Minor	30-40	Repeat every 12-24 hours	At least 1 day, until bleed stops	Moderate	30-40	Repeat every 12-24 hours	3+ days, until bleed stops	Major	35-50	Repeat every 12-24 hours	3+ days, until bleed stops	Life-Threatening	35-50	Repeat every 8-24 hours	Until threat has resolved
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VII. Billing Code/Availability Information

HCPCS Code & NDC:

Drug	Manufacturer	J-Code	1 Billable Unit Equiv.	Vial Size	NDC
Alphanate		J7186	1 IU	250 units	68516-4616-xx 68516-4611-xx

				500 units	68516-4617-xx 68516-4612-xx
				1000 units	68516-4618-xx 68516-4613-xx
				1500 units	68516-4619-xx 68516-4614-xx
				2000 units	68516-4620-xx 68516-4615-xx
	Grifols Biologicals LLC				
Humate-P	CSL Behring LLC	J7187	1 IU VWF:RCO	600 units	63833-0615-xx
				1200 units	63833-0616-xx
				2400 units	63833-0617-xx
Wilate	Octapharma USA Inc.	J7183	1 IU VWF:RCO	500 units	68982-0182-xx
				1000 units	

VIII. References

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3. Wilate [package insert]. Paramus, NJ; Octapharma USA Inc.; March 2020. Accessed May 2023.
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Appendix 1 – Covered Diagnosis Codes

ICD-10	ICD-10 Description
D66	Hereditary factor VIII deficiency
D68.01	Von Willebrand disease, type 1
D68.020	Von Willebrand disease, type 2A
D68.021	Von Willebrand disease, type 2B
D68.022	Von Willebrand disease, type 2M
D68.023	Von Willebrand disease, type 2N
D68.03	Von Willebrand disease, type 3
D68.04	Acquired von Willebrand disease
D68.09	Other von Willebrand disease

Appendix 2 – Centers for Medicare and Medicaid Services (CMS)

Medicare coverage for outpatient (Part B) drugs is outlined in the Medicare Benefit Policy Manual (Pub. 100-2), Chapter 15, §50 Drugs and Biologicals. In addition, National Coverage Determination (NCD), Local Coverage Determinations (LCDs), and Local Coverage Articles (LCAs) may exist and compliance with these policies is required where applicable. They can be found at: <https://www.cms.gov/medicare-coverage-database/search.aspx>. Additional indications may be covered at the discretion of the health plan.

Medicare Part B Covered Diagnosis Codes (applicable to existing NCD/LCD/LCA):

Jurisdiction(s): N	NCD/LCD Document (s): A56482
https://www.cms.gov/medicare-coverage-database/new-search/search-results.aspx?keyword=a56482&areaId=all&docType=NCA%2CCAL%2CNCD%2CMEDCAC%2CTA%2CMCD%2C6%2C3%2C5%2C1%2CF%2CP	

Jurisdiction(s): J,M	NCD/LCD Document (s): A56065
https://www.cms.gov/medicare-coverage-database/new-search/search-results.aspx?keyword=a56065&areaId=all&docType=NCA%2CCAL%2CNCD%2CMEDCAC%2CTA%2CMCD%2C6%2C3%2C5%2C1%2CF%2CP	

Jurisdiction(s): H,L	NCD/LCD Document (s): A56433
https://www.cms.gov/medicare-coverage-database/new-search/search-results.aspx?keyword=a56433&areaId=all&docType=NCA%2CCAL%2CNCD%2CMEDCAC%2CTA%2CMCD%2C6%2C3%2C5%2C1%2CF%2CP	

Medicare Part B Administrative Contractor (MAC) Jurisdictions		
Jurisdiction	Applicable State/US Territory	Contractor
E (1)	CA, HI, NV, AS, GU, CNMI	Noridian Healthcare Solutions, LLC
F (2 & 3)	AK, WA, OR, ID, ND, SD, MT, WY, UT, AZ	Noridian Healthcare Solutions, LLC
5	KS, NE, IA, MO	Wisconsin Physicians Service Insurance Corp (WPS)
6	MN, WI, IL	National Government Services, Inc. (NGS)
H (4 & 7)	LA, AR, MS, TX, OK, CO, NM	Novitas Solutions, Inc.
8	MI, IN	Wisconsin Physicians Service Insurance Corp (WPS)
N (9)	FL, PR, VI	First Coast Service Options, Inc.
J (10)	TN, GA, AL	Palmetto GBA, LLC
M (11)	NC, SC, WV, VA (excluding below)	Palmetto GBA, LLC
L (12)	DE, MD, PA, NJ, DC (includes Arlington & Fairfax counties and the city of Alexandria in VA)	Novitas Solutions, Inc.
K (13 & 14)	NY, CT, MA, RI, VT, ME, NH	National Government Services, Inc. (NGS)

Medicare Part B Administrative Contractor (MAC) Jurisdictions

Jurisdiction	Applicable State/US Territory	Contractor
15	KY, OH	CGS Administrators, LLC