

## SCIG (immune globulin SQ): Hizentra<sup>®</sup>, Gammagard Liquid<sup>®</sup>, Gamunex<sup>®</sup>-C, Gammaked<sup>®</sup>, Hyqvia<sup>®</sup>, Cuvitru<sup>®</sup>, Cutaquig<sup>®</sup>, Xembify<sup>®</sup> (Subcutaneous)

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### I. Length of Authorization

Initial coverage will be provided for 6 months and may be renewed annually thereafter.

### II. Dosing Limits

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

| Drug Name            | Dose/ week | Dose/28 days |
|----------------------|------------|--------------|
| Hizentra             | 46 g       | 184 g        |
| Gamunex-C & Gammaked | 24 g       | 96 g         |
| Gammagard liquid     | 24 g       | 96 g         |
| HyQvia               | 17.5 g     | 69 g         |
| Cuvitru              | 23 g       | 92 g         |
| Cutaquig             | 24 g       | 96 g         |
| Xembify              | 24 g       | 96 g         |

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

| Drug Name            | Billable units/28 days |
|----------------------|------------------------|
| Hizentra             | 960 (PID)              |
|                      | 1840 (CIDP)            |
| Gamunex-C & Gammaked | 192                    |
| Gammagard liquid     | 192                    |
| HyQvia               | 690                    |
| Cuvitru              | 920                    |
| Cutaquig             | N/A (96 gms/28 days)   |
| Xembify              | 960                    |

### III. Initial Approval Criteria

Baseline values for BUN and serum creatinine obtained within 30 days of request; **AND**

Coverage is provided in the following conditions:

#### **Primary immunodeficiency (PID)/Wiskott -Aldrich syndrome †**

Such as: x-linked agammaglobulinemia, common variable immunodeficiency, transient hypogammaglobulinemia of infancy, IgG subclass deficiency with or without IgA deficiency, antibody deficiency with near normal immunoglobulin levels) and combined deficiencies (severe combined immunodeficiencies, ataxia-telangiectasia, x-linked lymphoproliferative syndrome) [list not all inclusive]

- Patient has tried and failed to tolerate Intravenous Immunoglobulin (IVIG) therapy; **AND**
- Patient is  $\geq 2$  years old [*HyQvia and Cutaquig ONLY: patient must be  $\geq 18$  years old*]; **AND**
- Patient's IgG level is  $<200$  mg/dL **OR both** of the following
  - Patient has a history of multiple hard to treat infections as indicated by at least **one** of the following:
    - Four or more ear infections within 1 year
    - Two or more serious sinus infections within 1 year
    - Two or more months of antibiotics with little effect
    - Two or more pneumonias within 1 year
    - Recurrent or deep skin abscesses
    - Need for intravenous antibiotics to clear infections
    - Two or more deep-seated infections including septicemia; **AND**
  - The patient has a deficiency in producing antibodies in response to vaccination; **AND**
    - Titers were drawn before challenging with vaccination; **AND**
    - Titers were drawn between 4 and 8 weeks of vaccination

#### **Chronic Inflammatory Demyelinating Polyneuropathy (CIDP) [Hizentra ONLY] †**

- Patient must be  $\geq 18$  years old; **AND**
- Physician has assessed baseline disease severity utilizing an objective measure/tool (e.g., INCAT, Medical Research Council (MRC) muscle strength, 6-MWT, Rankin, Modified Rankin, etc.); **AND**
  - Used as initial maintenance therapy for prevention of disease relapses after treatment and stabilization with intravenous immunoglobulin (IVIG); **OR**
  - Used for re-initiation of maintenance therapy after experiencing a relapse and requiring re-induction therapy with IVIG (see Section IV for criteria)

#### **§ Initial IVIG criteria used for determination of coverage: (Reference Use Only)**

- Patient's disease course is progressive or relapsing and remitting for 2 months or longer; **AND**

**SCIG: Hizentra, Gammagard Liquid, Gamunex-C, Gammaked, Hyqvia, Cuvitru, Cutaquig, Xembify**  
**Prior Auth Criteria**

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- Patient has abnormal or absent deep tendon reflexes in upper or lower limbs; **AND**
- Electrodiagnostic testing indicating demyelination:
  - Partial motor conduction block in at least two motor nerves or in 1 nerve plus one other demyelination criterion listed here in at least 1 other nerve; **OR**
  - Distal CMAP duration increase in at least 1 nerve plus one other demyelination criterion listed here in at least 1 other nerve; **OR**
  - Abnormal temporal dispersion conduction must be present in at least 2 motor nerves; **OR**
  - Reduced conduction velocity in at least 2 motor nerves; **OR**
  - Prolonged distal motor latency in at least 2 motor nerves; **OR**
  - Absent F wave in at least two motor nerves plus one other demyelination criterion listed here in at least 1 other nerve; **OR**
  - Prolonged F wave latency in at least 2 motor nerves; **AND**
- Patient is refractory or intolerant to corticosteroids (e.g., prednisolone, prednisone, etc.) given in therapeutic doses over at least three months; **AND**
- Baseline in strength/weakness has been documented using an objective clinical measuring tool (e.g., INCAT, Medical Research Council (MRC) muscle strength, 6-MWT, Rankin, Modified Rankin, etc.)

† FDA Approved Indication(s)

#### IV. Renewal Criteria

Coverage can be renewed for 1 year based upon the following criteria:

- Patient continues to meet criteria identified in section III; **AND**
- Absence of unacceptable toxicity from the drug. Examples of unacceptable toxicity include: severe hypersensitivity/anaphylaxis, thrombosis, aseptic meningitis syndrome, hemolytic anemia, hyperproteinemia, acute lung injury, etc.; **AND**
- BUN and serum creatinine obtained within the last 6 months and the concentration and rate of infusion have been adjusted accordingly; **AND**

##### Primary immunodeficiency (PID)/Wiskott -Aldrich syndrome

- Disease response as evidenced by one or more of the following:
  - Decrease in the frequency of infection
  - Decrease in the severity of infection

##### Chronic Inflammatory Demyelinating Polyneuropathy (CIDP) [Hizentra ONLY]

- Renewals will be authorized for patients that have demonstrated a beneficial clinical response to maintenance therapy, without relapses, based on an objective clinical measuring tool (e.g., INCAT, Medical Research Council (MRC) muscle strength, 6-MWT, Rankin, Modified Rankin, etc.); **OR**
- Patient is re-initiating maintenance therapy after experiencing a relapse while on Hizentra; **AND**
  - Patient improved and stabilized on IVIG treatment; **AND**
  - Patient was NOT receiving maximum dosing of Hizentra prior to relapse

## V. Dosage/Administration

Dosing should be calculated using adjusted body weight if one or more of the following criteria are met:

- Patient's body mass index (BMI) is 30 kg/m<sup>2</sup> or more; **OR**
- Patient's actual body weight is 20% higher than his or her ideal body weight (IBW)

|  |
|--|
| Use the following dosing formulas to calculate the adjusted body weight (round dose to nearest 5 gram increment in adult patients) |
| <b>Dosing formulas</b>   |
| BMI = 703 x (weight in pounds/height in inches <sup>2</sup> )  |
| IBW(kg) for males = 50 + [2.3 (height in inches – 60)]   |
| IBW(kg) for females = 45.5 + [2.3 x (height in inches – 60)]   |
| Adjusted body weight = IBW + 0.5 (actual body weight – IBW)  |

*This information is not meant to replace clinical decision making when initiating or modifying medication therapy and should only be used as a guide. Patient-specific variables should be taken into account.*

| Indication   | Dose  |
|--|---|
| Chronic Inflammatory Demyelinating Polyneuropathy            | <p><u>Hizentra ONLY:</u></p> <ul style="list-style-type: none"> <li>▪ Initiate therapy 1 week after the last IVIG dose</li> <li>▪ The recommended subcutaneous dose is 0.2 g/kg (1 mL/kg) body weight per week, administered in 1 or 2 sessions over 1 or 2 consecutive days.</li> <li>▪ If CIDP symptoms worsen, consider re-initiating treatment with an IVIG while discontinuing Hizentra.               <ul style="list-style-type: none"> <li>– If improvement and stabilization are observed during IVIG treatment, consider reinitiating Hizentra at the dose of 0.4 g/kg body weight per week, administered in 2 sessions per week over 1 or 2 consecutive days, while discontinuing IVIG.</li> <li>– If CIDP symptoms worsen on the 0.4 g/kg body weight per week dose, consider re-initiating therapy with an IVIG while discontinuing Hizentra.</li> </ul> </li> </ul> |
| Primary immune deficiency including Wiskott-Aldrich Syndrome | <p><u>Hizentra:</u></p> <ul style="list-style-type: none"> <li>▪ Weekly dose: 1.37*(previous IVIG dose(g)/number of weeks between IVIG doses)</li> <li>▪ Biweekly dose: twice the weekly dose (using calculation above)</li> </ul>  |
|  | <p><u>Gamunex-C/Gammaked/Gammagard Liquid:</u></p> <ul style="list-style-type: none"> <li>▪ Weekly dose: 1.37*(previous IVIG dose(g)/number of weeks between IVIG doses)</li> </ul>   |
|  | <p><u>HyQvia:</u></p> <ul style="list-style-type: none"> <li>▪ Naïve to IgG or switching from SCIG: 300 to 600 mg/kg at 3 to 4 week intervals after initial ramp-up*</li> <li>▪ Switching from IGIV: use the same dose and frequency as the previous IV treatment after initial ramp-up*</li> </ul>   |

**SCIG: Hizentra, Gammagard Liquid, Gamunex-C, Gammaked, Hyqvia, Cuvitru, Cutaquig, Xembify**  
**Prior Auth Criteria**

| Indication | Dose  |
|------------|---|
|            | <p><u>Xembify:</u></p> <ul style="list-style-type: none"> <li>▪ Switching from IVIG : <ul style="list-style-type: none"> <li>○ Start treatment one week after the last IVIG infusion.</li> <li>○ Weekly dose: <math>1.37 \times (\text{previous monthly (or every 3- week) IVIG dose in grams}) / \text{number of weeks between IVIG doses}</math> <ul style="list-style-type: none"> <li>– To convert the dose in grams to mL, multiply the calculated initial SQ dose (in grams) by 5</li> <li>– Provided the total weekly dose is maintained, any dosing interval from daily up to weekly will achieve similar systemic IgG exposure when administered regularly at steady-state.</li> </ul> </li> </ul> </li> <li>▪ Switching from SCIG <ul style="list-style-type: none"> <li>○ Weekly dose (in grams) should be same as the weekly dose of prior SCIG treatment (in grams)</li> </ul> </li> </ul>   |
|            | <p><u>Cuvitru:</u></p> <ul style="list-style-type: none"> <li>▪ Switching from IVIG or HyQvia: <ul style="list-style-type: none"> <li>○ Weekly dose: <math>1.30 \times (\text{previous IVIG or HyQvia dose (g)}) / \text{number of weeks between IVIG or HyQvia doses}</math></li> <li>○ May be administered from daily up to every two weeks (biweekly)</li> <li>○ Biweekly dose: twice the weekly dose (using calculation above)</li> <li>○ Frequent dosing (2-7 times per week): divide the calculated weekly dose by the desired number of times per week</li> </ul> </li> <li>▪ Switching from SCIG <ul style="list-style-type: none"> <li>○ Weekly dose (in grams) should be same as the weekly dose of prior SCIG treatment (in grams)</li> <li>○ Biweekly dose: multiply the calculated weekly dose by 2</li> <li>○ Frequent dosing (2-7 times per week): divide the calculated weekly dose by the desired number of times per week</li> </ul> </li> </ul>                  |
|            | <p><u>Cutaquig:</u></p> <p><i>(Start treatment one week after the last IVIG or SCIG infusion. Ensure that patients have received IVIG or SCIG treatment at regular intervals for at least 3 months)</i></p> <ul style="list-style-type: none"> <li>▪ Switching from IVIG to Cutaquig <ul style="list-style-type: none"> <li>○ Establish the initial weekly dose by converting the monthly IVIG dose into an equivalent weekly dose and increasing it using a dose adjustment factor <ul style="list-style-type: none"> <li>– To calculate the initial weekly dose, divide the monthly IVIG dose in grams by the number of weeks between IGIV infusions and then multiply this value with a Dose Adjustment Factor of 1.40<br/><i>Initial weekly dose = Previous IGIV dose (in grams) x 1.40 / Number of weeks between IGIV doses</i></li> <li>– To convert the dose (in grams) to milliliters (mL), multiply the calculated dose (in grams) by 6</li> </ul> </li> </ul> </li> </ul> |

| Indication | Dose  |
|------------|---|
|            | <ul style="list-style-type: none"> <li>○ Provided the total weekly dose is maintained, any dosing interval from daily up to weekly can be used and will result in systemic IgG exposure that is comparable to the previous IGIV treatment</li> <li>▪ Switching from SCIG to Cutaquig <ul style="list-style-type: none"> <li>○ It is recommended to maintain the same weekly dosing (in grams) of Cutaquig that was used for the previous SCIG therapy (in grams) <ul style="list-style-type: none"> <li>– To convert the dose (in grams) to milliliters (mL), multiply the calculated dose (in grams) by 6</li> </ul> </li> <li>○ Obtain a trough IgG level before switching, monitor clinical response and check the trough IgG level 2 to 3 months after initiating Cutaquig</li> </ul> </li> </ul> |

*Dosing for immunoglobulin products is highly variable depending on numerous patient specific factors, indication(s), and the specific product selected. For specific dosing regimens refer to current prescribing literature.*

\*HyQvia initial treatment interval/dosage ramp-up schedule

| Week | Infusion Number          | 3-week treatment interval | 4-week treatment interval |
|------|--------------------------|---------------------------|---------------------------|
| 1    | 1 <sup>st</sup> infusion | Dose in Grams X 0.33      | Dose in Grams X 0.25      |
| 2    | 2 <sup>nd</sup> infusion | Dose in Grams X 0.67      | Dose in Grams X 0.50      |
| 4    | 3 <sup>rd</sup> infusion | Total Dose in Grams       | Dose in Grams X 0.75      |
| 7    | 4 <sup>th</sup> infusion | N/A                       | Total Dose in Grams       |

## VI. Billing Code/Availability Information

HCPSC code & NDC:

| Drug Name     | Manufacturer            | HCPSC Code  | 1 Billable unit | NDC           | IgG (grams) per SDV | Volume (mL) |
|---------------|-------------------------|---|-----------------|---------------|---------------------|-------------|
| Hizentra 20%  | CSL Behring AG          | J1559 – Injection, immune globulin (Hizentra), 100 mg   | 100 mg          | 44206-0451-01 | 1                   | 5           |
|               |                         |   |                 | 44206-0452-02 | 2                   | 10          |
|               |                         |   |                 | 44206-0454-04 | 4                   | 20          |
|               |                         |   |                 | 44206-0455-10 | 10                  | 50          |
| Gammaked 10%  | Kedrion Biopharma, Inc. | J1561 Injection, immune globulin, (Gamunex-C/Gammaked), non-lyophilized (e.g. liquid), 500 mg   | 500 mg          | 76125-0900-01 | 1                   | 10          |
|               |                         |   |                 | 76125-0900-25 | 2.5                 | 25          |
|               |                         |   |                 | 76125-0900-50 | 5                   | 50          |
|               |                         |   |                 | 76125-0900-10 | 10                  | 100         |
| Gamunex-C 10% | Grifols Therapeutics    | J1561 – Injection, immune globulin, (Gamunex-C/Gammaked), non-lyophilized (e.g. liquid), 500 mg | 500 mg          | 13533-0800-12 | 1                   | 10          |
|               |                         |   |                 | 13533-0800-15 | 2.5                 | 25          |
|               |                         |   |                 | 13533-0800-20 | 5                   | 50          |
|               |                         |   |                 | 13533-0800-71 | 10                  | 100         |
|               |                         |   |                 | 13533-0800-24 | 20                  | 200         |
|               |                         |   |                 | 13533-0800-40 | 40                  | 400         |

**SCIG: Hizentra, Gammagard Liquid, Gamunex-C, Gammaked, Hyqvia, Cuvitru, Cutaquig, Xembify**  
**Prior Auth Criteria**

| Drug Name  | Manufacturer                  | HCP Code   | 1 Billable unit | NDC           | IgG (grams) per SDV | Volume (mL) |
|--|-------------------------------|--|-----------------|---------------|---------------------|-------------|
| Gammagard Liquid 10%                                       | Baxter Healthcare Corporation | J1569 – Injection, immune globulin, (Gammagard liquid), non-lyophilized, (e.g. liquid), 500 mg | 500 mg          | 00944-2700-02 | 1                   | 10          |
|  |                               |  |                 | 00944-2700-03 | 2.5                 | 25          |
|  |                               |  |                 | 00944-2700-04 | 5                   | 50          |
|  |                               |  |                 | 00944-2700-05 | 10                  | 100         |
|  |                               |  |                 | 00944-2700-06 | 20                  | 200         |
|  |                               |  |                 | 00944-2700-07 | 30                  | 300         |
| HyQvia 10% (with Recombinant Human Hyaluronidase 160 U/mL) | Baxter Healthcare Corporation | J1575 – Injection, immune globulin/hyaluronidase, (Hyqvia), 100 mg immune globulin             | 100 mg          | 00944-2510-02 | 2.5                 | 25          |
|  |                               |  |                 | 00944-2511-02 | 5                   | 50          |
|  |                               |  |                 | 00944-2512-02 | 10                  | 100         |
|  |                               |  |                 | 00944-2513-02 | 20                  | 200         |
| Cuvitru 20%  | Baxalta US Inc.               | J1555 – Injection, immune globulin (Cuvitru), 100 mg   | 100 mg          | 00944-2850-01 | 1                   | 5           |
|  |                               |  |                 | 00944-2850-03 | 2                   | 10          |
|  |                               |  |                 | 00944-2850-05 | 4                   | 20          |
|  |                               |  |                 | 00944-2850-07 | 8                   | 40          |
| Cutaquig 16.5%   | Octapharma                    | J3590; 90284   | N/A             | 68892-0810-01 | 1                   | 6           |
|  |                               |  |                 | 68892-0810-02 | 1.65                | 10          |
|  |                               |  |                 | 68892-0810-03 | 2                   | 12          |
|  |                               |  |                 | 68892-0810-04 | 3.3                 | 20          |
|  |                               |  |                 | 68892-0810-05 | 4                   | 24          |
|  |                               |  |                 | 68892-0810-06 | 8                   | 48          |
| Xembify 20%  | Grifols                       | J3590; 90284; J1558*   | N/A             | 13533-0810-05 | 1                   | 5           |
|  |                               |  |                 | 13533-0810-10 | 2                   | 10          |
|  |                               |  |                 | 13533-0810-20 | 4                   | 20          |
|  |                               |  |                 | 13533-0810-50 | 10                  | 50          |
| Immune Globulin, Human, Subcutaneous                       | N/A                           | J3590 – unclassified biologic; C9399 – unclassified drug or biological                         | N/A             | N/A           | N/A                 | N/A         |
|  |                               | 90284 – immune globulin (SCIg), human, for use in subcutaneous infusions                       |                 |               |                     |             |

\*Effective 07/01/2020

## VII. References

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SCIg: Hizentra, Gammagard Liquid, Gamunex-C, Gammaked, Hyqvia, Cuvitru, Cutaquig, Xembify  
Prior Auth Criteria

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20. Wisconsin Physicians Service Insurance Corporation. Local Coverage Determination (LCD): Immune Globulins (L34771). Centers for Medicare & Medicaid Services, Inc. Updated on 07/23/2019 with effective date 8/13/2019. Accessed August 2019.

## Appendix 1 – Covered Diagnosis Codes

| ICD-10 | ICD-10 Description   |
|--------|--|
| B20    | Human immunodeficiency virus [HIV] disease   |
| D80.0  | Hereditary hypogammaglobulinemia   |
| D80.1  | Nonfamilial hypogammaglobulinemia  |
| D80.2  | Selective deficiency of immunoglobulin A [IgA]   |
| D80.3  | Selective deficiency of immunoglobulin G [IgG] subclasses                                      |
| D80.4  | Selective deficiency of immunoglobulin M [IgM]   |
| D80.5  | Immunodeficiency with increased immunoglobulin M [IgM]   |
| D80.7  | Transient hypogammaglobulinemia of infancy   |
| D81.0  | Severe combined immunodeficiency [SCID] with reticular dysgenesis                              |
| D81.1  | Severe combined immunodeficiency [SCID] with low T- and B-cell numbers                         |
| D81.2  | Severe combined immunodeficiency [SCID] with low or normal B-cell numbers                      |
| D81.6  | Major histocompatibility complex class I deficiency  |
| D81.7  | Major histocompatibility complex class II deficiency   |
| D81.89 | Other combined immunodeficiencies  |
| D81.9  | Combined immunodeficiency, unspecified   |
| D82.0  | Wiskott-Aldrich syndrome   |
| D83.0  | Common variable immunodeficiency with predominant abnormalities of B-cell numbers and function |
| D83.2  | Common variable immunodeficiency with autoantibodies to B- or T-cells                          |
| D83.8  | Other common variable immunodeficiencies   |
| D83.9  | Common variable immunodeficiency, unspecified  |
| G61.81 | Chronic inflammatory demyelinating polyneuritis  |
| G61.89 | Other inflammatory polyneuropathies  |
| G62.89 | Other specified polyneuropathies   |

## 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) and Local Coverage Determinations (LCDs) may exist and compliance with these policies is required where applicable. They can be found at: <http://www.cms.gov/medicare-coverage-database/search/advanced-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):

|   |   |
|---|---|
| <b>Jurisdiction(s):</b> N   | <b>NCD/LCD/Article Document (s):</b> L34007 |
| <a href="https://www.cms.gov/medicare-coverage-database/search/lcd-date-search.aspx?DocID=L34007&amp;bc=gAAAAAAAAAAAAA==">https://www.cms.gov/medicare-coverage-database/search/lcd-date-search.aspx?DocID=L34007&amp;bc=gAAAAAAAAAAAAA==</a> |   |

|   |   |
|---|---|
| <b>Jurisdiction(s):</b> 5, 8  | <b>NCD/LCD/Article Document (s):</b> L34771 |
| <a href="https://www.cms.gov/medicare-coverage-database/search/lcd-date-search.aspx?DocID=L34771&amp;bc=gAAAAAAAAAAAAA==">https://www.cms.gov/medicare-coverage-database/search/lcd-date-search.aspx?DocID=L34771&amp;bc=gAAAAAAAAAAAAA==</a> |   |

| <b>Medicare Part B Administrative Contractor (MAC) Jurisdictions</b> |   |   |
|--|---|---|
| <b>Jurisdiction</b>  | <b>Applicable State/US Territory</b>  | <b>Contractor</b>                                 |
| 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)          |
| 15   | KY, OH  | CGS Administrators, LLC                           |