PB 132 of 2023

National Health (Highly Specialised Drugs Program) Special Arrangement Amendment (January Update) Instrument 2023

National Health Act 1953

I, NIKOLAI TSYGANOV, Assistant Secretary, Pricing and PBS Policy Branch, Technology Assessment and Access Division, Department of Health and Aged Care, delegate of the Minister for Health and Aged Care, make this Instrument under subsection 100(2) of the *National Health Act 1953*.

Dated 21 December 2023

NIKOLAI TSYGANOV
Assistant Secretary
Pricing and PBS Policy Branch
Technology Assessment and Access Division

Contents

1 Name

(1) This instrument is the *National Health (Highly Specialised Drugs Program) Special Arrangement Amendment (January Update) Instrument 2023*.

(2) This instrument may also be cited as PB 132 of 2023.

2 Commencement

(1) Each provision of this instrument specified in column 1 of the table commences, or is taken to have commenced, in accordance with column 2 of the table. Any other statement in column 2 has effect according to its terms.

Commencement information		
Column 1	**Column 2**	**Column 3**
Provisions	**Commencement**	**Date/Details**
1. *The whole of this instrument*	*1 January 2024*	*1 January 2024*

Note: This table relates only to the provisions of this instrument as originally made. It will not be amended to deal with any later amendments of this instrument.

(2) Any information in column 3 of the table is not part of this instrument. Information may be inserted in this column, or information in it may be edited, in any published version of this instrument.

3 Authority

This instrument is made under subsection 100(2) of the *National Health Act 1953*.

4 Schedules

Each instrument that is specified in a Schedule to this instrument is amended or repealed as set out in the applicable items in the Schedule concerned, and any other item in a Schedule to this instrument has effect according to its terms.

Schedule 1—Amendments

National Health (Highly Specialised Drugs Program) Special Arrangement 2021 (PB 27 of 2021)

[1] Schedule 1, entry for Abacavir with Lamivudine

insert in the columns in the order indicated, and in alphabetical order for the column headed "Brand":

		Abacavir/Lamivudine Viatris	C4527 C4528		60	5

[2] Schedule 1, entry for Adalimumab in each of the forms: Injection 40 mg in 0.4 mL pre-filled pen; and Injection 40 mg in 0.4 mL pre-filled syringe

insert in the columns in the order indicated, and in alphabetical order for the column headed "Brand":

		Adalicip	C12120 C14061 C14063 C14064 C14107 C14136		See Schedule 2	See Schedule 2

[3] Schedule 1, entry for Deferasirox in the form Tablet, dispersible, 125 mg

substitute:

Tablet, dispersible, 125 mg	Oral	Deferasirox Juno	C7374 C7375 C7385 C8326 C8328 C8329 C9222 C9258 C9302	P7385 P8326 P8328 P8329 P9222 P9258 P9302	168	2
		Pharmacor Deferasirox	C7374 C7375 C7385 C8326 C8328 C8329 C9222 C9258 C9302	P7385 P8326 P8328 P8329 P9222 P9258 P9302	168	2
		Deferasirox Juno	C7374 C7375 C7385 C8326 C8328 C8329 C9222 C9258 C9302	P7374 P7375	168	5

		Pharmacor Deferasirox	C7374 C7375 C7385 C8326 C8328 C8329 C9222 C9258 C9302	P7374 P7375	168	5

[4] Schedule 1, entry for Deferasirox in the form Tablet, dispersible, 250 mg

substitute:

Tablet, dispersible, 250 mg	Oral	Deferasirox Juno	C7374 C7375 C7385 C8326 C8328 C8329 C9222 C9258 C9302	P7385 P8326 P8328 P8329 P9222 P9258 P9302	168	2
		Pharmacor Deferasirox	C7374 C7375 C7385 C8326 C8328 C8329 C9222 C9258 C9302	P7385 P8326 P8328 P8329 P9222 P9258 P9302	168	2
		Deferasirox Juno	C7374 C7375 C7385 C8326 C8328 C8329 C9222 C9258 C9302	P7374 P7375	168	5
		Pharmacor Deferasirox	C7374 C7375 C7385 C8326 C8328 C8329 C9222 C9258 C9302	P7374 P7375	168	5

[5] Schedule 1, entry for Deferasirox in the form Tablet, dispersible, 500 mg

substitute:

Tablet, dispersible, 500 mg	Oral	Deferasirox Juno	C7374 C7375 C7385 C8326 C8328 C8329 C9222 C9258 C9302	P7385 P8326 P8328 P8329 P9222 P9258 P9302	168	2

			Pharmacor Deferasirox	C7374 C7375 C7385 C8326 C8328 C8329 C9222 C9258 C9302	P7385 P8326 P8328 P8329 P9222 P9258 P9302	168	2
			Deferasirox Juno	C7374 C7375 C7385 C8326 C8328 C8329 C9222 C9258 C9302	P7374 P7375	168	5
			Pharmacor Deferasirox	C7374 C7375 C7385 C8326 C8328 C8329 C9222 C9258 C9302	P7374 P7375	168	5

[6] Schedule 1, entry for Eculizumab

substitute:

Eculizumab	Solution concentrate for I.V. infusion 300 mg in 30 mL	Injection	Soliris	C13458 C13459 C13464 C13560 C13660 C13661 C13684 C13845 C13857 C14750 C14753 C14754 C14781 C14792 C14793 C14799 C14805	See Schedule 2	See Schedule 2

[7] Schedule 1, entry for Lumacaftor with ivacaftor

substitute:

Lumacaftor with ivacaftor	Sachet containing granules, lumacaftor 75 mg and ivacaftor 94 mg	Oral	Orkambi	C14757 C14765	See Schedule 2	See Schedule 2
	Sachet containing granules, lumacaftor 100 mg and ivacaftor 125 mg	Oral	Orkambi	C14757 C14765	See Schedule 2	See Schedule 2
	Sachet containing granules, lumacaftor 150 mg	Oral	Orkambi	C14757 C14765	See Schedule 2	See Schedule 2

	and ivacaftor 188 mg					
	Tablet containing lumacaftor 100 mg with ivacaftor 125 mg	Oral	Orkambi	C14783 C14784	See Schedule 2	See Schedule 2
	Tablet containing lumacaftor 200 mg with ivacaftor 125 mg	Oral	Orkambi	C14785 C14796	See Schedule 2	See Schedule 2

[8] Schedule 1, entry for Pomalidomide

substitute:

Pomalidomide	Capsule 1 mg	Oral	Pomolide	C13746 C13755 C13757 C13768	See Schedule 2	See Schedule 2
	Capsule 2 mg	Oral	Pomolide	C13746 C13755 C13757 C13768	See Schedule 2	See Schedule 2
	Capsule 3 mg	Oral	Pomalidomide Sandoz	C13746 C13755 C13757 C13768	See Schedule 2	See Schedule 2
			Pomalyst	C13746 C13755 C13757 C13768	See Schedule 2	See Schedule 2
			Pomolide	C13746 C13755 C13757 C13768	See Schedule 2	See Schedule 2
	Capsule 4 mg	Oral	Pomalidomide Sandoz	C13746 C13755 C13757 C13768	See Schedule 2	See Schedule 2
			Pomalyst	C13746 C13755 C13757 C13768	See Schedule 2	See Schedule 2
			Pomolide	C13746 C13755 C13757 C13768	See Schedule 2	See Schedule 2

[9] Schedule 1, entry for Ravulizumab in each of the forms: Solution concentrate for I.V. infusion 300 mg in 3 mL; and Solution concentrate for I.V. infusion 1,100 mg in 11 mL

insert in numerical order in the column headed "Circumstances": **C14744 C14746 C14747 C14748 C14749 C14780 C14791 C14797**

[10] Schedule 1, entry for Ustekinumab

insert in numerical order in the column headed "Circumstances": **C14758 C14787 C14801**

[11] Schedule 2, entry for Eculizumab

substitute:

Eculizumab	C14781	Sufficient for treatment for 4 weeks	0
	C13857	1	0
	C14750 C14792	Sufficient for treatment for 4 weeks	4
	C14753 C14754 C14793 C14799 C14805	Sufficient for treatment for 4 weeks	5
	C13464 C13660 C13661 C13684 C13845	6	5
	C13458 C13459 C13560	8	0

[12] Schedule 2, entry for Lumacaftor with Ivacaftor

substitute:

Lumacaftor with ivacaftor	C14757 C14765 C14783 C14784 C14785 C14796	1 pack	5

[13] Schedule 2, entry for Pomalidomide *[Maximum Quantity: 1 pack (21 capsules); Maximum Repeats: 5]*

omit from the column headed "Maximum Repeats": **5** *substitute:* **0**

[14] Schedule 2, entry for Ravulizumab

substitute:

Ravulizumab	C13459 C14477 C14565 C14586 C14744 C14780 C14791 C14797	1 dose	0
	C14476 C14530 C14531 C14746 C14747 C14748 C14749	1 dose	2

[15] Schedule 2, entry for Ustekinumab

insert in numerical order in the column headed "Circumstances": **C14758 C14787 C14801**

[16] Schedule 3, entry for Eculizumab

(a) *omit:*

	C6626		Atypical haemolytic uraemic syndrome (aHUS) Initial treatment Patient must have active and progressing thrombotic microangiopathy (TMA) caused by aHUS; AND Patient must have ADAMTS-13 activity of greater than or equal to 10% on a blood sample taken	Compliance with Written Authority Required procedures

			prior to plasma exchange or infusion; or, if ADAMTS-13 activity was not collected prior to plasma exchange or infusion, patient must have platelet counts of greater than 30x10^9/L and a serum creatinine of greater than 150 mol/L; AND Patient must have a confirmed negative STEC (Shiga toxin-producing E.Coli) result if the patient has had diarrhoea in the preceding 14 days; AND Patient must have clinical features of active organ damage or impairment; AND Patient must not receive more than 4 weeks of treatment under this restriction. Must be treated by a paediatric nephrologist, a nephrologist, a paediatric haematologist or a haematologist, or, must be in consultation with a paediatric nephrologist, a nephrologist, a paediatric haematologist or a haematologist. Evidence of active and progressing TMA is defined by the following: (1) a platelet count of less than 150x10^9/L; and evidence of two of the following: (i) presence of schistocytes on blood film; (ii) low or absent haptoglobin; (iii) lactate dehydrogenase (LDH) above normal range; OR (2) in recipients of a kidney transplant for end-stage kidney disease due to aHUS, a kidney biopsy confirming TMA; AND (3) evidence of at least one of the following clinical features of active TMA-related organ damage or impairment is defined as below: (a) kidney impairment as demonstrated by one of the following: (i) a decline in estimated Glomerular Filtration Rate (eGFR) of greater than 20% in a patient who has pre-existing kidney impairment; and/or (ii) a serum creatinine (sCr) of greater than the upper limit of normal (ULN) in a patient who has no history of pre-existing kidney impairment; or (iii) a sCr of greater than the age-appropriate ULN in paediatric patients; or (iv) a renal biopsy consistent with aHUS; (b) onset of TMA-related neurological impairment; (c) onset of TMA-related cardiac impairment; (d) onset of TMA-related gastrointestinal impairment; (e) onset of TMA-related pulmonary impairment. Claims of non-renal TMA-related organ damage should be made at the point of application for initial PBS-subsidised eculizumab (where possible), and should be supported by objective clinical measures. The prescriber's cover letter should establish that the observed organ damage is directly linked to active and progressing TMA, particularly when indirect causes such as severe thrombocytopenia, hypertension and acute renal failure are present at the time of the initial organ impairment. Serial haematological results (every 3 months while the patient is receiving treatment) must be provided with every subsequent application for treatment. The authority application must be in writing and must include: (1) A completed authority prescription form; and (2) A completed aHUS eculizumab Authority Application Supporting Information Form - Initial PBS-subsidised eculizumab treatment; and	

			(3) A signed patient acknowledgement or an acknowledgement signed by a parent or authorised guardian, if applicable; and (4) A detailed cover letter from the prescriber; and (5) A copy of a current Certificate of vaccination or a statement that vaccination has or will be administered and appropriate antibiotic prophylaxis has been prescribed; and (6) A measurement of body weight at the time of application; and (7) The result of ADAMTS-13 activity on a blood sample taken prior to plasma exchange or infusion; the date and time that the sample for the ADAMTS-13 assay was collected, and the dates and times of any plasma exchanges or infusions that were undertaken in the two weeks prior to collection of the ADAMTS-13 assay; and (8) In the case that a sample for ADAMTS-13 assay was not collected prior to plasma exchange or infusion, measurement of ADAMTS-13 activity must be taken 1-2 weeks following the last plasma exchange or infusion. The ADAMTS-13 result must be submitted to the Department of Human Services within 27 days of commencement of eculizumab treatment in order for the patient to be considered as eligible for further PBS-subsidised eculizumab treatment, underInitial treatment 1-balance of supply; and (9) A confirmed negative STEC result if the patient has had diarrhoea in the preceding 14 days; and (10) Evidence of active and progressing TMA, including pathology results where relevant. Evidence of the onset of TMA-related neurological, cardiac, gastrointestinal or pulmonary impairment requires a supporting statement with clinical evidence in patient records. All tests must have been performed within one month of application; and (11) For all patients, a recent measurement of eGFR, platelets and two of either LDH, haptoglobin or schistocytes of no more than 1 week old at the time of application.	
	C6637		Atypical haemolytic uraemic syndrome (aHUS) Extended initial treatment - Assessment phase Patient must have received treatment under the initial restriction with PBS subsidised eculizumab for this condition; AND Patient must have demonstrated on-going treatment response of PBS-subsidised eculizumab treatment for this condition; AND Patient must not have experienced treatment failure with eculizumab including PBS-subsidised eculizumab for this condition; AND Patient must not receive more than 56 weeks of treatment under this restriction. Must be treated by a paediatric nephrologist, a nephrologist, a paediatric haematologist or a haematologist, or, must be in consultation with a paediatric nephrologist, a nephrologist, a paediatric haematologist or a haematologist. A treatment response is defined as: (1) Normalisation of haematology as demonstrated by at least 2 of the following: platelet count, haptoglobin, and LDH; AND (2) One of the following: a) An increase in eGFR of > 25% from baseline, where the baseline is the eGFR measurement immediately prior to commencing treatment with eculizumab or b) an eGFR within +/- 25% from baseline; or	Compliance with Written Authority Required procedures

			c) an avoidance of dialysis-dependence but worsening of kidney function with a reduction in eGFR 25% from baseline. PBS-subsidised treatment with eculizumab will not be permitted if a patient has experienced treatment failure. A treatment failure is defined as a patient who is: (1) dialysis-dependent at the time of application and has failed to demonstrate significant resolution of extra-renal complications if originally presented; or (2) on dialysis and has been on dialysis for 4 months of the previous 6 months while receiving PBS-subsidised eculizumab and has failed to demonstrate significant resolution of extra-renal complications if originally presented. A maximum of up to 56 weeks of treatment is allowed under this restriction, however an application must be submitted at 6 months, 12 months, 18 months and 24 months following commencing PBS-subsidised eculizumab. The authority application must include the following measures of response to the prior course of treatment, including serial haematological results (every 3 months while the patient is receiving treatment). The authority application must be in writing and must include: (1) A completed authority prescription form; and (2) A completed aHUS eculizumab Authority Application Supporting Information Form for Extended Initial treatment; and (3) A detailed cover letter from the prescriber; and (4) A copy of a current Certificate of vaccination or a statement that vaccination has or will be administered and appropriate antibiotic prophylaxis has been prescribed; and (5) A measurement of body weight at the time of application; and (6) An identified genetic mutation, if applicable; and (7) A family history of aHUS, if applicable; and (8) A history of multiple episodes of aHUS before commencing eculizumab treatment, if applicable; and (9) A history of kidney transplant, if applicable, (especially if required due to aHUS); and (10) An inclusion of the individual consequences of recurrent disease, if applicable; and (11) Evidence that the patient has had a treatment response including haematological results of no more than 1 week old at the time of application (platelet count, haptoglobin and LDH); and an eGFR level of no more than 1 week old at the time of application; and (12) Evidence that the patient has not experienced treatment failure, including a supporting statement with clinical evidence that the patient does not require dialysis, unless the indication for continuing eculizumab is severe extra-renal complications that have significantly improved; and (13) If the indication for continuing eculizumab is severe extra-renal complications, then a supporting statement with clinical evidence that any initial extra-renal complications of TMA have significantly improved is required. This assessment must be submitted no later than 4 weeks from the cessation of the prior treatment. Where a response assessment is not undertaken and submitted within these timeframes, the patient will be deemed to have failed to respond to treatment with eculizumab.	
	C6642		Atypical haemolytic uraemic syndrome (aHUS)	Compliance with Written

			Initial treatment - Balance of Supply Must be treated by a paediatric nephrologist, a nephrologist, a paediatric haematologist or a haematologist, or, must be in consultation with a paediatric nephrologist, a nephrologist, a paediatric haematologist or a haematologist. Patient must have received PBS-subsidised initial supply of eculizumab for this condition; AND Patient must have ADAMTS-13 activity of greater than or equal to 10% on a blood sample; AND Patient must not receive more than 20 weeks supply under this restriction. ADAMTS-13 activity result must have been submitted to the Department of Human Services. In the case that a sample for ADAMTS-13 activity taken prior to plasma exchange or infusion was not available at the time of application for Initial Treatment, ADAMTS-13 activity must have been measured 1-2 weeks following the last plasma exchange or infusion, and must have been submitted to the Department of Human Services within 27 days of commencement of eculizumab. The date and time that the sample for the ADAMTS-13 assay was collected, and the dates and times of the last, if any, plasma exchange or infusion that was undertaken in the two weeks prior to collection of the ADAMTS-13 assay must also have been provided to Department of Human Services. Serial haematological results (every 3 months while the patient is receiving treatment) must be provided with every subsequent application for treatment.	Authority Required procedures
	C6668		Atypical haemolytic uraemic syndrome (aHUS) Continuing treatment Patient must have received treatment under Extended Initial restriction with PBS subsidised eculizumab for this condition; AND Patient must have demonstrated on-going treatment response of PBS-subsidised eculizumab treatment for this condition; AND Patient must not have experienced treatment failure with eculizumab including PBS-subsidised eculizumab for this condition; AND Patient must not receive more than 24 weeks of treatment under this restriction. Must be treated by a paediatric nephrologist, a nephrologist, a paediatric haematologist or a haematologist, or, must be in consultation with a paediatric nephrologist, a nephrologist, a paediatric haematologist or a haematologist. A treatment response is defined as: (1) Normalisation of haematology as demonstrated by at least 2 of the following: platelet count, haptoglobin, and LDH; AND (2) One of the following: a) An increase in eGFR of > 25% from baseline, where the baseline is the eGFR measurement immediately prior to commencing treatment with eculizumab or b) an eGFR within +/- 25% from baseline; or c) an avoidance of dialysis-dependence but worsening of kidney function with a reduction in eGFR 25% from baseline. PBS-subsidised treatment with eculizumab will not be permitted if a patient has experienced treatment failure. A treatment failure is defined as a patient who is: (1) dialysis-dependent at the time of application and has failed to demonstrate significant	Compliance with Written Authority Required procedures

			resolution of extra-renal complications if originally presented; or (2) on dialysis and has been on dialysis for 4 months of the previous 6 months while receiving PBS-subsidised eculizumab and has failed to demonstrate significant resolution of extra-renal complications if originally presented. The authority application must include the following measures of response to the prior course of treatment, including serial haematological results (every 3 months while the patient is receiving treatment). The authority application must be in writing and must include: (1) A completed authority prescription form; and (2) A completed aHUS eculizumab Authority Application Supporting Information Form for Continuing treatment; and (3) A detailed cover letter from the prescriber; and (4) A copy of a current Certificate of vaccination or a statement that vaccination has or will be administered and appropriate antibiotic prophylaxis has been prescribed; and (5) A measurement of body weight at the time of application; and (6) An identified genetic mutation, if applicable; and (7) A family history of aHUS, if applicable; and (8) A history of multiple episodes of aHUS before recommencing eculizumab treatment, if applicable; and (9) A history of kidney transplant if applicable (especially if required due to aHUS); and (10) An inclusion of the individual consequences of recurrent disease, if applicable; and (11) Evidence that the patient has had a treatment response including haematological results of no more than 1 week old at the time of application (platelet count, haptoglobin and LDH); and an eGFR level of no more than 1 week old at the time of application; and (12) Evidence that the patient has not experienced treatment failure, including a supporting statement with clinical evidence that the patient does not require dialysis, unless the indication for continuing eculizumab is severe extra-renal complications that have significantly improved; and (13) If the indication for continuing eculizumab is severe extra-renal complications, then a supporting statement with clinical evidence that any initial extra-renal complications of TMA have significantly improved is required. This assessment must be submitted no later than 4 weeks from the cessation of the prior treatment. Where a response assessment is not undertaken and submitted within these timeframes, the patient will be deemed to have failed to respond to treatment with eculizumab.	
	C6686		Atypical haemolytic uraemic syndrome (aHUS) Extended Continuing treatment Patient must have received treatment under the Continuing treatment with PBS-subsidised eculizumab for this condition; AND Patient must have demonstrated on-going treatment response with PBS-subsidised eculizumab for this condition; AND Patient must not have ever experienced treatment failure with eculizumab including PBS-subsidised eculizumab for this condition; AND Patient must have a TMA-related cardiomyopathy as evidenced by left ventricular ejection fraction < 40% on current objective measurement; OR	Compliance with Written Authority Required procedures

			Patient must have severe TMA-related neurological impairment; OR Patient must have severe TMA-related gastrointestinal impairment; OR Patient must have severe TMA-related pulmonary impairment on current objective measurement; OR Patient must have grade 4 or 5 chronic kidney disease (eGFR of less than 30 mL/min); OR Patient must have a high risk of aHUS recurrence in the short term in the absence of continued treatment with eculizumab; AND Patient must not receive more than 24 weeks of treatment per continuing treatment course authorised under this restriction. Must be treated by a paediatric nephrologist, a nephrologist, a paediatric haematologist or a haematologist, or, must be in consultation with a paediatric nephrologist, a nephrologist, a paediatric haematologist or a haematologist. A treatment response is defined as: (1) Normalisation of haematology as demonstrated by at least 2 of the following: platelet count, haptoglobin, and LDH; AND (2) One of the following: a) An increase in eGFR of > 25% from baseline, where the baseline is the eGFR measurement immediately prior to commencing treatment with eculizumab or b) an eGFR within +/- 25% from baseline; or c) an avoidance of dialysis-dependence but worsening of kidney function with a reduction in eGFR 25% from baseline. PBS-subsidised treatment with eculizumab will not be permitted if a patient has experienced treatment failure. A treatment failure is defined as a patient who is: (1) dialysis-dependent at the time of application and has failed to demonstrate significant resolution of extra-renal complications if originally presented; or (2) on dialysis and has been on dialysis for 4 months of the previous 6 months while receiving PBS-subsidised eculizumab and has failed to demonstrate significant resolution of extra-renal complications if originally presented. The authority application must include the following measures of response to the prior course of treatment, including serial haematological results (every 3 months while the patient is receiving treatment). The authority application must be in writing and must include: (1) A completed authority prescription form; and (2) A completed aHUS eculizumab Authority Application Supporting Information Form for Continuing treatment; and (3) A detailed cover letter from the prescriber; and (4) A copy of a current Certificate of vaccination or a statement that vaccination has or will be administered and appropriate antibiotic prophylaxis has been prescribed; and (5) A measurement of body weight at the time of application; and (6) An identified genetic mutation, if applicable; and (7) A family history of aHUS, if applicable; and (8) A history of multiple episodes of aHUS before commencing eculizumab treatment, if applicable; and (9) A history of kidney transplant, if applicable (especially if required due to aHUS); and	

			(10) An inclusion of the individual consequences of recurrent disease; and (11) A supporting statement with clinical evidence of severe TMA-related cardiomyopathy (including current LVEF result), neurological impairment, gastrointestinal impairment or pulmonary impairment; and (12) Evidence that the patient has had a treatment response including haematological results of no more than 1 month old at the time of application (platelet count, haptoglobin and LDH); and an eGFR level of no more than 1 month old at the time of application; and (13) Evidence that the patient has not experienced treatment failure, including a supporting statement with clinical evidence that the patient does not require dialysis, unless the indication for continuing eculizumab is severe extra-renal complications that have significantly improved; and (14) If the indication for continuing eculizumab is severe extra-renal complications, then a supporting statement with clinical evidence that any initial extra-renal complications of TMA have significantly improved is required. This assessment must be submitted no later than 4 weeks from the cessation of the prior treatment. Where a response assessment is not undertaken and submitted within these timeframes, the patient will be deemed to have failed to respond to treatment with eculizumab.	
	C6687		Atypical haemolytic uraemic syndrome (aHUS) Recommencement of treatment Patient must have demonstrated treatment response to previous treatment with PBS-subsidised eculizumab for this condition; AND Patient must not have ever experienced treatment failure with eculizumab including PBS-subsidised eculizumab for this condition; AND Patient must have the following clinical conditions:(i) either significant haemolysis as measured by low/absent haptoglobin; or presence of schistocytes on the blood film; or lactate dehydrogenase (LDH) above normal;AND(ii) either platelet consumption as measured by either 25% decline from patient baseline or thrombocytopenia (platelet count <150 x 10^9/L);OR(iii) TMA-related organ impairment including on recent biopsy; AND Patient must not receive more than 24 weeks of treatment under this restriction. Must be treated by a paediatric nephrologist, a nephrologist, a paediatric haematologist or a haematologist, or, must be in consultation with a paediatric nephrologist, a nephrologist, a paediatric haematologist or a haematologist. A treatment response is defined as: (1) Normalisation of haematology as demonstrated by at least 2 of the following: platelet count, haptoglobin, and LDH; AND (2) One of the following: a) An increase in eGFR of > 25% from baseline, where the baseline is the eGFR measurement immediately prior to commencing treatment with eculizumab or b) an eGFR within +/- 25% from baseline; or c) an avoidance of dialysis-dependence but worsening of kidney function with a reduction in eGFR 25% from baseline. PBS-subsidised treatment with eculizumab will not be permitted if a patient has experienced treatment failure. A treatment failure is defined as a patient who is: (1) dialysis-dependent at the time of application and has failed to demonstrate significant	Compliance with Written Authority Required procedures

			resolution of extra-renal complications if originally presented; or (2) on dialysis and has been on dialysis for 4 months of the previous 6 months while receiving PBS-subsidised eculizumab and has failed to demonstrate significant resolution of extra-renal complications if originally presented. The authority application must include the following measures of response to the prior course of treatment, including serial haematological results (every 3 months while the patient is receiving treatment). The authority application must be in writing and must include: (1) A completed authority prescription form(s); and (2) A completed aHUS eculizumab Authority Application Supporting Information Form for Recommencement of treatment; and (3) A signed patient acknowledgement or an acknowledgement signed by a parent or authorised guardian, if applicable; and (4) A detailed cover letter from the prescriber; and (5) A copy of a current Certificate of vaccination or a statement that vaccination has or will be administered and appropriate antibiotic prophylaxis has been prescribed; and (6) A measurement of body weight at the time of application, and (7) An identified genetic mutation, if applicable; and (8) A family history of aHUS if applicable; and (9) A history of multiple episodes of aHUS following the treatment break, if applicable; and (10) A history of kidney transplant if applicable (especially if required due to aHUS); and (11) An inclusion of the individual consequences of recurrent disease; and (12) A supporting statement with clinical evidence of TMA-related organ damage including current (within one week of application) haematological results (platelet count, haptoglobin and LDH), eGFR level, and, if applicable, on recent biopsy; (13) Evidence that the patient has had a treatment response to their previous treatment with eculizumab; and (14) Evidence that the patient has not experienced treatment failure, including a supporting statement with clinical evidence that the patient does not require dialysis, unless the indication for continuing eculizumab is severe extra-renal complications that have significantly improved; and (15) If the indication for continuing eculizumab is severe extra-renal complications, then a supporting statement with clinical evidence that any initial extra-renal complications of TMA have significantly improved is required. This assessment must be submitted no later than 4 weeks from the cessation of the prior treatment. Where a response assessment is not undertaken and submitted within these timeframes, the patient will be deemed to have failed to respond to treatment with eculizumab.	
	C6688		Atypical haemolytic uraemic syndrome (aHUS) Continuing recommencement of treatment Patient must have received treatment under Recommencement of treatment restriction with PBS-subsidised eculizumab for this condition; AND Patient must have demonstrated ongoing treatment response to the previous 24 weeks of PBS-subsidised eculizumab for this condition; AND Patient must not have experienced treatment failure with eculizumab including PBS-subsidised	Compliance with Written Authority Required procedures

			eculizumab for this condition; AND Patient must not receive more than 24 weeks of treatment under this restriction. Must be treated by a paediatric nephrologist, a nephrologist, a paediatric haematologist or a haematologist, or, must be in consultation with a paediatric nephrologist, a nephrologist, a paediatric haematologist or a haematologist. A treatment response is defined as: (1) Normalisation of haematology as demonstrated by at least 2 of the following: platelet count, haptoglobin, and LDH; AND (2) One of the following: a) An increase in eGFR of > 25% from baseline, where the baseline is the eGFR measurement immediately prior to commencing treatment with eculizumab or b) an eGFR within +/- 25% from baseline; or c) an avoidance of dialysis-dependence but worsening of kidney function with a reduction in eGFR 25% from baseline. PBS-subsidised treatment with eculizumab will not be permitted if a patient has experienced treatment failure. A treatment failure is defined as a patient who is: (1) dialysis-dependent at the time of application and has failed to demonstrate significant resolution of extra-renal complications if originally presented; or (2) on dialysis and has been on dialysis for 4 months of the previous 6 months while receiving PBS-subsidised eculizumab and has failed to demonstrate significant resolution of extra-renal complications if originally presented. The authority application must include the following measures of response to the prior course of treatment, including serial haematological results (every 3 months while the patient is receiving treatment). The authority application must be in writing and must include: (1) A completed authority prescription form; and (2) A completed aHUS eculizumab Authority Application Supporting Information Form for Continuing treatment; and (3) A detailed cover letter from the prescriber; and (4) A copy of a current Certificate of vaccination or a statement that vaccination has or will be administered and appropriate antibiotic prophylaxis has been prescribed; and (5) A measurement of body weight at the time of application; and (6) An identified genetic mutation, if applicable; and (7) A family history of aHUS, if applicable; and (8) A history of multiple episodes of aHUS before recommencing eculizumab treatment, if applicable; and (9) A history of kidney transplant if applicable (especially if required due to aHUS); and (10) An inclusion of the individual consequences of recurrent disease, if applicable; and (11) Evidence that the patient has had a treatment response including haematological results of no more than 1 week old at the time of application (platelet count, haptoglobin and LDH); and an eGFR level of no more than 1 week old at the time of application; and (12) Evidence that the patient has not experienced treatment failure, including a supporting statement with clinical evidence that the patient does not require dialysis, unless the indication for continuing eculizumab is severe extra-renal complications that have significantly improved; and	

			(13) If the indication for continuing eculizumab is severe extra-renal complications, then a supporting statement with clinical evidence that any initial extra-renal complications of TMA have significantly improved is required. This assessment must be submitted no later than 4 weeks from the cessation of the prior treatment. Where a response assessment is not undertaken and submitted within these timeframes, the patient will be deemed to have failed to respond to treatment with eculizumab.	

(b) *insert in numerical order after existing text:*

	C14750		Atypical haemolytic uraemic syndrome (aHUS) Recommencement - Balance of Supply Patient must have previously received PBS-subsidised eculizumab under the 'Recommencement of treatment' restriction for this condition; AND Patient must not receive more than 20 weeks supply under this restriction. Must be treated by a prescriber who is either: (i) a haematologist, (ii) a nephrologist; OR Must be treated by a medical practitioner who has consulted at least one of the above mentioned specialist types, with agreement reached that the patient should be treated with this pharmaceutical benefit on this occasion; AND Patient must be undergoing treatment with one C5 inhibitor therapy only at any given time. Serial haematological results (every 3 months while the patient is receiving treatment) must be provided with every subsequent application for treatment.	Compliance with Written Authority Required procedures
	C14753		Atypical haemolytic uraemic syndrome (aHUS) Switch from PBS-subsidised ravulizumab (all phases) - loading dose Patient must have previously received PBS-subsidised ravulizumab under the 'Initial treatment' restriction for this condition; OR Patient must have previously received PBS-subsidised ravulizumab under the 'Continuing treatment' restriction for this condition; OR Patient must have previously received PBS-subsidised ravulizumab under the 'Extended continuing treatment' restriction for this condition; OR Patient must have previously received PBS-subsidised ravulizumab under the 'Recommencement of treatment' restriction for this condition; OR Patient must have previously received PBS-subsidised ravulizumab under the 'Continuing recommencement of treatment' restriction for this condition; OR Patient must have previously received PBS-subsidised ravulizumab under the 'Grandfather (transitioning from non-PBS to PBS-subsidised treatment)' restriction for this condition; AND Patient must have/had ADAMTS-13 activity of greater than or equal to 10% on a blood sample; AND Patient must not receive more than 24 weeks of C5 inhibitor supply for this current treatment phase under this restriction. Must be treated by a prescriber who is either: (i) a haematologist, (ii) a nephrologist; OR Must be treated by a medical practitioner who has consulted at least one of the above mentioned specialist types, with agreement reached that the patient should be treated with this pharmaceutical benefit on this occasion; AND	Compliance with Written Authority Required procedures

			Patient must be undergoing treatment with one C5 inhibitor therapy only at any given time. The application must indicate the most recent treatment phase that the patient is switching from. For patients who are switching C5 inhibitors, the next application should be sought under the next relevant treatment phase. Serial haematological results (every 3 months while the patient is receiving treatment) must be provided with every subsequent application for treatment. The authority application must be in writing and must include all of the following: (1) A completed authority prescription form(s); (2) A completed authority application form relevant to the indication and treatment phase (the latest version is located on the website specified in the Administrative Advice); (3) A measurement of body weight at the time of application; (4) Results of genetic testing, if not previously submitted.	
	C14754		Atypical haemolytic uraemic syndrome (aHUS) Continuing treatment Patient must have received PBS-subsidised eculizumab under the initial treatment phase for this condition; OR Patient must have received PBS-subsidised eculizumab under the switch from ravulizumab in the initial treatment phase for this condition; OR Patient must have received PBS-subsidised eculizumab under the switch from ravulizumab in the continuing treatment phase for this condition; AND Patient must have demonstrated on-going treatment response with PBS-subsidised eculizumab for this condition; AND Patient must not have experienced treatment failure with eculizumab for this condition in the most recent treatment phase; AND Patient must not receive more than 80 weeks of eculizumab treatment in total under this restriction; OR Patient must not receive more than 104 weeks supply of a C5 inhibitor under the initial and continuing treatment restrictions if they had switched C5 inhibitors during the course of initial and continuing treatment; AND Patient must not receive more than 24 weeks of treatment per continuing treatment course authorised under this restriction. Must be treated by a prescriber who is either: (i) a haematologist, (ii) a nephrologist; OR Must be treated by a medical practitioner who has consulted at least one of the above mentioned specialist types, with agreement reached that the patient should be treated with this pharmaceutical benefit on this occasion; AND Patient must be undergoing treatment with one C5 inhibitor therapy only at any given time. A treatment response is defined as: (1) Normalisation of haematology as demonstrated by at least 2 of the following: (i) platelet count, (ii) haptoglobin, (iii) lactate dehydrogenase (LDH); and (2) One of the following: a) an increase in eGFR of > 25% from baseline, where the baseline is the eGFR measurement immediately prior to commencing treatment with a C5 inhibitor; or b) an eGFR within +/- 25% from baseline; or	Compliance with Written Authority Required procedures

			c) an avoidance of dialysis-dependence but worsening of kidney function with a reduction in eGFR 25% from baseline. PBS-subsidised treatment with eculizumab will not be permitted if a patient has experienced treatment failure with eculizumab in the most recent treatment phase prior to the treatment phase where this application is sought. A treatment failure is defined as a patient who is: (1) Dialysis-dependent at the time of application and has failed to demonstrate significant resolution of extra-renal complications if originally presented; or (2) On dialysis and has been on dialysis for 4 months of the previous 6 months while receiving a PBS-subsidised C5 inhibitor, and has failed to demonstrate significant resolution of extra-renal complications if originally presented. The authority application must include the following measures of response to the prior course of treatment, including serial haematological results (every 3 months while the patient is receiving treatment). The authority application must be in writing and must include all of the following: (1) A completed authority prescription form(s); (2) A completed authority application form relevant to the indication and treatment phase (the latest version is located on the website specified in the Administrative Advice); (3) A measurement of body weight at the time of application; (4) Results of genetic testing, if not previously submitted; (5) A family history of aHUS, if applicable; (6) A history of kidney transplant if applicable (especially if required due to aHUS); (7) An inclusion of the individual consequences of recurrent disease, if applicable; (8) Evidence that the patient has had a treatment response including haematological results of no more than 1 week old at the time of application (platelet count, haptoglobin and LDH); and an eGFR level of no more than 1 week old at the time of application; (9) Evidence that the patient has not experienced treatment failure, including a supporting statement with clinical evidence that the patient does not require dialysis, unless the indication for continuing eculizumab is severe extra-renal complications that have significantly improved; (10) If the indication for continuing eculizumab is severe extra-renal complications, then a supporting statement with clinical evidence that any initial extra-renal complications of TMA have significantly improved is required. This assessment must be submitted no later than 4 weeks from the cessation of the prior treatment. Where a response assessment is not undertaken and submitted within these timeframes, the patient will be deemed to have failed to respond to treatment with eculizumab.	
	C14781		Atypical haemolytic uraemic syndrome (aHUS) Initial treatment Patient must have active and progressing thrombotic microangiopathy (TMA) caused by aHUS; AND Patient must have ADAMTS-13 activity of greater than or equal to 10% on a blood sample taken prior to plasma exchange or infusion; or, if ADAMTS-13 activity was not collected prior to plasma exchange or infusion, patient must have platelet counts of greater than 30x10^9/L and a serum creatinine of greater than 150 mol/L; AND	Compliance with Written Authority Required procedures

			Patient must have a confirmed negative STEC (Shiga toxin-producing E.Coli) result if the patient has had diarrhoea in the preceding 14 days; AND Patient must have clinical features of active organ damage or impairment; AND Patient must not receive more than 4 weeks of treatment under this restriction. Must be treated by a prescriber who is either: (i) a haematologist, (ii) a nephrologist; OR Must be treated by a medical practitioner who has consulted at least one of the above mentioned specialist types, with agreement reached that the patient should be treated with this pharmaceutical benefit on this occasion; AND Patient must be undergoing treatment with one C5 inhibitor therapy only at any given time. Evidence of active and progressing TMA is defined by the following: (1) a platelet count of less than 150x10^9/L; and evidence of two of the following: (i) presence of schistocytes on blood film; (ii) low or absent haptoglobin; (iii) lactate dehydrogenase (LDH) above normal range; OR (2) in recipients of a kidney transplant for end-stage kidney disease due to aHUS, a kidney biopsy confirming TMA; AND (3) evidence of at least one of the following clinical features of active TMA-related organ damage or impairment is defined as below: (a) kidney impairment as demonstrated by one of the following: (i) a decline in estimated Glomerular Filtration Rate (eGFR) of greater than 20% in a patient who has pre-existing kidney impairment; and/or (ii) a serum creatinine (sCr) of greater than the upper limit of normal (ULN) in a patient who has no history of pre-existing kidney impairment; or (iii) a sCr of greater than the age-appropriate ULN in paediatric patients; or (iv) a renal biopsy consistent with aHUS; (b) onset of TMA-related neurological impairment; (c) onset of TMA-related cardiac impairment; (d) onset of TMA-related gastrointestinal impairment; (e) onset of TMA-related pulmonary impairment. Claims of non-renal TMA-related organ damage should be made at the point of application for initial PBS-subsidised eculizumab (where possible), and should be supported by objective clinical measures. The prescriber's cover letter should establish that the observed organ damage is directly linked to active and progressing TMA, particularly when indirect causes such as severe thrombocytopenia, hypertension and acute renal failure are present at the time of the initial organ impairment. Serial haematological results (every 3 months while the patient is receiving treatment) must be provided with every subsequent application for treatment. The authority application must be in writing and must include all of the following: (1) A completed authority prescription form(s); (2) A completed authority application form relevant to the indication and treatment phase (the latest version is located on the website specified in the Administrative Advice); (3) A detailed cover letter from the prescriber;	

			(4) A measurement of body weight at the time of application; (5) The result of ADAMTS-13 activity on a blood sample taken prior to plasma exchange or infusion; the date and time that the sample for the ADAMTS-13 assay was collected, and the dates and times of any plasma exchanges or infusions that were undertaken in the 2 weeks prior to collection of the ADAMTS-13 assay; (6) In the case that a sample for ADAMTS-13 assay was not collected prior to plasma exchange or infusion, measurement of ADAMTS-13 activity must be taken 7-10 days following the last plasma exchange or infusion. The ADAMTS-13 result must be submitted to Services Australia within 27 days of commencement of eculizumab treatment in order for the patient to be considered as eligible for further PBS-subsidised eculizumab treatment, under Initial treatment - Balance of Supply; (7) A confirmed negative STEC result if the patient has had diarrhoea in the preceding 14 days; (8) Evidence of active and progressing TMA, including pathology results where relevant. Evidence of the onset of TMA-related neurological, cardiac, gastrointestinal or pulmonary impairment requires a supporting statement with clinical evidence in patient records. All tests must have been performed within 4 weeks of application; (9) For all patients, a recent measurement of eGFR, platelets and two of either LDH, haptoglobin or schistocytes of no more than 1 week old at the time of application.	
	C14792		Atypical haemolytic uraemic syndrome (aHUS) Initial treatment - Balance of Supply Patient must have received PBS-subsidised initial supply of eculizumab for this condition; AND Patient must have ADAMTS-13 activity of greater than or equal to 10% on a blood sample; AND Patient must not receive more than 20 weeks supply under this restriction. Must be treated by a prescriber who is either: (i) a haematologist, (ii) a nephrologist; OR Must be treated by a medical practitioner who has consulted at least one of the above mentioned specialist types, with agreement reached that the patient should be treated with this pharmaceutical benefit on this occasion; AND Patient must be undergoing treatment with one C5 inhibitor therapy only at any given time. ADAMTS-13 activity result must have been submitted to Services Australia. In the case that a sample for ADAMTS-13 activity taken prior to plasma exchange or infusion was not available at the time of application for Initial treatment, ADAMTS-13 activity must have been measured 7-10 days following the last plasma exchange or infusion, and must have been submitted to Services Australia within 27 days of commencement of eculizumab. The date and time that the sample for the ADAMTS-13 assay was collected, and the dates and times of the last, if any, plasma exchange or infusion that was undertaken in the 2 weeks prior to collection of the ADAMTS-13 assay must also have been provided to Services Australia. Serial haematological results (every 3 months while the patient is receiving treatment) must be provided with every subsequent application for treatment.	Compliance with Written Authority Required procedures
	C14793		Atypical haemolytic uraemic syndrome (aHUS) Continuing recommencement of treatment Patient must have received PBS-subsidised eculizumab under the recommencement of treatment phase for this condition; OR	Compliance with Written Authority Required procedures

			Patient must have received PBS-subsidised eculizumab under the switch from ravulizumab in the recommencement treatment phase for this condition; OR Patient must have received PBS-subsidised eculizumab under the switch from ravulizumab in the continuing recommencement of treatment phase for this condition; AND Patient must have demonstrated ongoing treatment response to 'Recommencement of treatment' with a C5 inhibitor for this condition; AND Patient must not have experienced treatment failure with eculizumab for this condition in the most recent treatment phase; AND Patient must not receive more than 24 weeks of treatment with eculizumab per continuing treatment course authorised under this restriction. Must be treated by a prescriber who is either: (i) a haematologist, (ii) a nephrologist; OR Must be treated by a medical practitioner who has consulted at least one of the above mentioned specialist types, with agreement reached that the patient should be treated with this pharmaceutical benefit on this occasion; AND Patient must be undergoing treatment with one C5 inhibitor therapy only at any given time. A treatment response is defined as: (1) Normalisation of haematology as demonstrated by at least 2 of the following: (i) platelet count, (ii) haptoglobin, (iii) lactate dehydrogenase (LDH); and (2) One of the following: a) an increase in eGFR of > 25% from baseline, where the baseline is the eGFR measurement immediately prior to commencing treatment with a C5 inhibitor; or b) an eGFR within +/- 25% from baseline; or c) an avoidance of dialysis-dependence but worsening of kidney function with a reduction in eGFR 25% from baseline. PBS-subsidised treatment with eculizumab will not be permitted if a patient has experienced treatment failure with eculizumab in the most recent treatment phase prior to the treatment phase where this application is sought. A treatment failure is defined as a patient who is: (1) Dialysis-dependent at the time of application and has failed to demonstrate significant resolution of extra-renal complications if originally presented; or (2) On dialysis and has been on dialysis for 4 months of the previous 6 months while receiving a PBS-subsidised C5 inhibitor, and has failed to demonstrate significant resolution of extra-renal complications if originally presented. The authority application must include the following measures of response to the prior course of treatment, including serial haematological results (every 3 months while the patient is receiving treatment). The authority application must be in writing and must include all of the following: (1) A completed authority prescription form(s); (2) A completed authority application form relevant to the indication and treatment phase (the latest version is located on the website specified in the Administrative Advice); (3) A measurement of body weight at the time of application; (4) Results of genetic testing, if not previously submitted; (5) A family history of aHUS, if applicable; (6) A history of multiple episodes of aHUS before recommencing eculizumab treatment, if	

			applicable; (7) A history of kidney transplant if applicable (especially if required due to aHUS); (8) An inclusion of the individual consequences of recurrent disease, if applicable; (9) Evidence that the patient has had a treatment response including haematological results of no more than 1 week old at the time of application (platelet count, haptoglobin and LDH); and an eGFR level of no more than 1 week old at the time of application; (10) Evidence that the patient has not experienced treatment failure, including a supporting statement with clinical evidence that the patient does not require dialysis, unless the indication for continuing eculizumab is severe extra-renal complications that have significantly improved; (11) If the indication for continuing eculizumab is severe extra-renal complications, then a supporting statement with clinical evidence that any initial extra-renal complications of TMA have significantly improved is required. This assessment must be submitted no later than 4 weeks from the cessation of the prior treatment. Where a response assessment is not undertaken and submitted within these timeframes, the patient will be deemed to have failed to respond to treatment with eculizumab.	
	C14799		Atypical haemolytic uraemic syndrome (aHUS) Recommencement of treatment Patient must have demonstrated treatment response to previous treatment with PBS-subsidised eculizumab for this condition; OR Patient must have received PBS-subsidised eculizumab under the switch from ravulizumab in the recommencement treatment phase for this condition; AND Patient must not have experienced treatment failure with eculizumab for this condition in the most recent treatment phase; AND Patient must have the following clinical conditions prior to recommencing C5 inhibitor treatment: (i) either significant haemolysis as measured by low/absent haptoglobin; or presence of schistocytes on the blood film; or lactate dehydrogenase (LDH) above normal; AND (ii) either platelet consumption as measured by either 25% decline from patient baseline or thrombocytopenia (platelet count <150 x 10^9/L); OR (iii) TMA-related organ impairment including on recent biopsy; AND Patient must not receive more than 24 weeks of treatment under this restriction. Must be treated by a prescriber who is either: (i) a haematologist, (ii) a nephrologist; OR Must be treated by a medical practitioner who has consulted at least one of the above mentioned specialist types, with agreement reached that the patient should be treated with this pharmaceutical benefit on this occasion; AND Patient must be undergoing treatment with one C5 inhibitor therapy only at any given time. A treatment response is defined as: (1) Normalisation of haematology as demonstrated by at least 2 of the following: (i) platelet count, (ii) haptoglobin, (iii) lactate dehydrogenase (LDH); and (2) One of the following: a) an increase in eGFR of > 25% from baseline, where the baseline is the eGFR measurement immediately prior to commencing treatment with a C5 inhibitor; or b) an eGFR within +/- 25% from baseline; or c) an avoidance of dialysis-dependence but worsening of kidney function with a reduction in eGFR	Compliance with Written Authority Required procedures

			25% from baseline. PBS-subsidised treatment with eculizumab will not be permitted if a patient has experienced treatment failure with eculizumab in the most recent treatment phase prior to the treatment phase where this application is sought. A treatment failure is defined as a patient who is: (1) Dialysis-dependent at the time of application and has failed to demonstrate significant resolution of extra-renal complications if originally presented; or (2) On dialysis and has been on dialysis for 4 months of the previous 6 months while receiving a PBS-subsidised C5 inhibitor, and has failed to demonstrate significant resolution of extra-renal complications if originally presented. The authority application must include the following measures of response to the prior course of treatment, including serial haematological results (every 3 months while the patient is receiving treatment). The authority application must be in writing and must include all of the following: (1) A completed authority prescription form(s); (2) A completed authority application form relevant to the indication and treatment phase (the latest version is located on the website specified in the Administrative Advice); (3) A measurement of body weight at the time of application; (4) Results of genetic testing, if not previously submitted; (5) A family history of aHUS if applicable; (6) A history of multiple episodes of aHUS following the treatment break, if applicable; (7) A history of kidney transplant if applicable (especially if required due to aHUS); (8) An inclusion of the individual consequences of recurrent disease; (9) A supporting statement with clinical evidence of TMA-related organ damage including current (within one week of application) haematological results (platelet count, haptoglobin and LDH), eGFR level, and, if applicable, on recent biopsy; (10) Evidence that the patient has had a treatment response to their previous treatment with eculizumab; (11) Evidence that the patient has not experienced treatment failure, including a supporting statement with clinical evidence that the patient does not require dialysis, unless the indication for continuing eculizumab is severe extra-renal complications that have significantly improved; (12) If the indication for continuing eculizumab is severe extra-renal complications, then a supporting statement with clinical evidence that any initial extra-renal complications of TMA have significantly improved is required.	
	C14805		Atypical haemolytic uraemic syndrome (aHUS) Extended Continuing treatment Patient must have received PBS-subsidised eculizumab under the continuing treatment phase for this condition; OR Patient must have received PBS-subsidised eculizumab under the switch from ravulizumab in the continuing treatment phase for this condition; OR Patient must have received PBS-subsidised eculizumab under the switch from ravulizumab in the extended continuing treatment phase for this condition; AND Patient must have demonstrated on-going treatment response with PBS-subsidised eculizumab	Compliance with Written Authority Required procedures

			for this condition; AND Patient must not have experienced treatment failure with eculizumab for this condition in the most recent treatment phase; AND Patient must have a TMA-related cardiomyopathy as evidenced by left ventricular ejection fraction < 40% on current objective measurement; OR Patient must have severe TMA-related neurological impairment; OR Patient must have severe TMA-related gastrointestinal impairment; OR Patient must have severe TMA-related pulmonary impairment on current objective measurement; OR Patient must have grade 4 or 5 chronic kidney disease (eGFR of less than 30 mL/min); OR Patient must have a high risk of aHUS recurrence in the short term in the absence of continued treatment with eculizumab; AND Patient must not receive more than 24 weeks of treatment per continuing treatment course authorised under this restriction. Must be treated by a prescriber who is either: (i) a haematologist, (ii) a nephrologist; OR Must be treated by a medical practitioner who has consulted at least one of the above mentioned specialist types, with agreement reached that the patient should be treated with this pharmaceutical benefit on this occasion; AND Patient must be undergoing treatment with one C5 inhibitor therapy only at any given time. A treatment response is defined as: (1) Normalisation of haematology as demonstrated by at least 2 of the following: (i) platelet count, (ii) haptoglobin, (iii) lactate dehydrogenase (LDH); and (2) One of the following: a) an increase in eGFR of > 25% from baseline, where the baseline is the eGFR measurement immediately prior to commencing treatment with a C5 inhibitor; or b) an eGFR within +/- 25% from baseline; or c) an avoidance of dialysis-dependence but worsening of kidney function with a reduction in eGFR 25% from baseline. PBS-subsidised treatment with eculizumab will not be permitted if a patient has experienced treatment failure with eculizumab in the most recent treatment phase prior to the treatment phase where this application is sought. A treatment failure is defined as a patient who is: (1) Dialysis-dependent at the time of application and has failed to demonstrate significant resolution of extra-renal complications if originally presented; or (2) On dialysis and has been on dialysis for 4 months of the previous 6 months while receiving a PBS-subsidised C5 inhibitor, and has failed to demonstrate significant resolution of extra-renal complications if originally presented. The authority application must include the following measures of response to the prior course of treatment, including serial haematological results (every 3 months while the patient is receiving treatment). The authority application must be in writing and must include all of the following: (1) A completed authority prescription form(s); (2) A completed authority application form relevant to the indication and treatment phase (the latest version is located on the website specified in the Administrative Advice);	

			(3) A measurement of body weight at the time of application; (4) Results of genetic testing, if not previously submitted; (5) A family history of aHUS, if applicable; (6) A history of multiple episodes of aHUS before commencing eculizumab treatment, if applicable; (7) A history of kidney transplant, if applicable (especially if required due to aHUS); (8) An inclusion of the individual consequences of recurrent disease; (9) A supporting statement with clinical evidence of severe TMA-related cardiomyopathy (including current LVEF result), neurological impairment, gastrointestinal impairment or pulmonary impairment; (10) Evidence that the patient has had a treatment response including haematological results of no more than 4 weeks old at the time of application (platelet count, haptoglobin and LDH); and an eGFR level of no more than 4 weeks old at the time of application; (11) Evidence that the patient has not experienced treatment failure, including a supporting statement with clinical evidence that the patient does not require dialysis, unless the indication for continuing eculizumab is severe extra-renal complications that have significantly improved; (12) If the indication for continuing eculizumab is severe extra-renal complications, then a supporting statement with clinical evidence that any initial extra-renal complications of TMA have significantly improved is required. This assessment must be submitted no later than 4 weeks from the cessation of the prior treatment. Where a response assessment is not undertaken and submitted within these timeframes, the patient will be deemed to have failed to respond to treatment with eculizumab.	

[17] Schedule 3, entry for Lumacaftor with ivacaftor

substitute:

Lumacaftor with ivacaftor	C14757		Cystic fibrosis Continuing treatment Must be treated by a specialist respiratory physician with expertise in cystic fibrosis or in consultation with a specialist respiratory physician with expertise in cystic fibrosis if attendance is not possible due to geographic isolation; AND Must be treated in a centre with expertise in cystic fibrosis or in consultation with a centre with expertise in cystic fibrosis if attendance is not possible due to geographic isolation. Patient must have previously received PBS-subsidised treatment with this drug for this condition; AND The treatment must be the sole PBS-subsidised cystic fibrosis transmembrane conductance regulator (CFTR) modulator therapy for this condition; AND The treatment must be given concomitantly with standard therapy for this condition. Patient must be 1 year of age or older. This pharmaceutical benefit is not PBS-subsidised for this condition in a patient who is currently receiving one of the strong CYP3A4 inducers outlined in the Product Information. The authority application must be in writing and must include: (1) a completed authority prescription; and	Compliance with Written Authority Required procedures

			(2) a completed Cystic Fibrosis Continuing Authority Application Supporting Information Form; and (3) current CYP3A4 inhibitors, CYP3A4 inducers and IV antibiotics.	
	C14765		Cystic fibrosis Initial treatment Must be treated by a specialist respiratory physician with expertise in cystic fibrosis or in consultation with a specialist respiratory physician with expertise in cystic fibrosis if attendance is not possible due to geographic isolation; AND Must be treated in a centre with expertise in cystic fibrosis or in consultation with a centre with expertise in cystic fibrosis if attendance is not possible due to geographic isolation. Patient must be homozygous for the F508del mutation in the cystic fibrosis transmembrane conductance regulator (CFTR) gene; AND The treatment must be given concomitantly with standard therapy for this condition; AND The treatment must be the sole PBS-subsidised cystic fibrosis transmembrane conductance regulator (CFTR) modulator therapy for this condition. Patient must be 1 year of age or older. This pharmaceutical benefit is not PBS-subsidised for this condition in a patient who is currently receiving one of the strong CYP3A4 inducers outlined in the Product Information. The authority application must be in writing and must include: (1) a completed authority prescription; and (2) a completed Cystic Fibrosis Authority Application Supporting Information Form; and (3) details of the pathology report substantiating the patient being homozygous for the F508del mutation on the CFTR gene - quote each of the: (i) name of the pathology report provider, (ii) date of pathology report, (iii) unique identifying number/code that links the pathology result to the individual patient; and (4) current CYP3A4 inhibitors, CYP3A4 inducers and IV antibiotics.	Compliance with Written Authority Required procedures
	C14783		Cystic fibrosis Initial treatment Must be treated by a specialist respiratory physician with expertise in cystic fibrosis or in consultation with a specialist respiratory physician with expertise in cystic fibrosis if attendance is not possible due to geographic isolation; AND Must be treated in a centre with expertise in cystic fibrosis or in consultation with a centre with expertise in cystic fibrosis if attendance is not possible due to geographic isolation. Patient must be homozygous for the F508del mutation in the cystic fibrosis transmembrane conductance regulator (CFTR) gene; AND The treatment must be given concomitantly with standard therapy for this condition; AND Patient must have either chronic sinopulmonary disease or gastrointestinal and nutritional abnormalities; AND The treatment must be the sole PBS-subsidised cystic fibrosis transmembrane conductance regulator (CFTR) modulator therapy for this condition. Patient must be aged between 6 and 11 years inclusive. This pharmaceutical benefit is not PBS-subsidised for this condition in a patient who is currently receiving one of the strong CYP3A4 inducers outlined in the Product Information.	Compliance with Written Authority Required procedures

			The authority application must be in writing and must include: (1) a completed authority prescription; and (2) a completed Cystic Fibrosis Authority Application Supporting Information Form; and (3) details of the pathology report substantiating the patient being homozygous for the F508del mutation on the CFTR gene - quote each of the: (i) name of the pathology report provider, (ii) date of pathology report, (iii) unique identifying number/code that links the pathology result to the individual patient; and (4) current CYP3A4 inhibitors, CYP3A4 inducers and IV antibiotics.	
	C14784		Cystic fibrosis Continuing treatment Must be treated by a specialist respiratory physician with expertise in cystic fibrosis or in consultation with a specialist respiratory physician with expertise in cystic fibrosis if attendance is not possible due to geographic isolation; AND Must be treated in a centre with expertise in cystic fibrosis or in consultation with a centre with expertise in cystic fibrosis if attendance is not possible due to geographic isolation. Patient must have previously received PBS-subsidised treatment with this drug for this condition; AND The treatment must be the sole PBS-subsidised cystic fibrosis transmembrane conductance regulator (CFTR) modulator therapy for this condition; AND The treatment must be given concomitantly with standard therapy for this condition. Patient must be aged between 6 and 11 years inclusive. This pharmaceutical benefit is not PBS-subsidised for this condition in a patient who is currently receiving one of the strong CYP3A4 inducers outlined in the Product Information. The authority application must be in writing and must include: (1) a completed authority prescription; and (2) a completed Cystic Fibrosis Continuing Authority Application Supporting Information Form; and (3) current CYP3A4 inhibitors, CYP3A4 inducers and IV antibiotics.	Compliance with Written Authority Required procedures
	C14785		Cystic fibrosis Continuing treatment Must be treated by a specialist respiratory physician with expertise in cystic fibrosis or in consultation with a specialist respiratory physician with expertise in cystic fibrosis if attendance is not possible due to geographic isolation; AND Must be treated in a centre with expertise in cystic fibrosis or in consultation with a centre with expertise in cystic fibrosis if attendance is not possible due to geographic isolation. Patient must have previously received PBS-subsidised treatment with this drug for this condition; AND The treatment must be given concomitantly with standard therapy for this condition; AND The treatment must be the sole PBS-subsidised cystic fibrosis transmembrane conductance regulator (CFTR) modulator therapy for this condition. Patient must be 12 years of age or older. This pharmaceutical benefit is not PBS-subsidised for this condition in a patient who is currently receiving one of the strong CYP3A4 inducers outlined in the Product Information.	Compliance with Written Authority Required procedures

			The authority application must be in writing and must include: (1) a completed authority prescription; and (2) a completed Cystic Fibrosis Continuing Authority Application Supporting Information Form; and (3) current CYP3A4 inhibitors, CYP3A4 inducers and IV antibiotics.	
	C14796		Cystic fibrosis Initial treatment Must be treated by a specialist respiratory physician with expertise in cystic fibrosis or in consultation with a specialist respiratory physician with expertise in cystic fibrosis if attendance is not possible due to geographic isolation; AND Must be treated in a centre with expertise in cystic fibrosis or in consultation with a centre with expertise in cystic fibrosis if attendance is not possible due to geographic isolation. Patient must be homozygous for the F508del mutation in the cystic fibrosis transmembrane conductance regulator (CFTR) gene; AND The treatment must be given concomitantly with standard therapy for this condition; AND Patient must have either chronic sinopulmonary disease or gastrointestinal and nutritional abnormalities; AND The treatment must be the sole PBS-subsidised cystic fibrosis transmembrane conductance regulator (CFTR) modulator therapy for this condition. Patient must be 12 years of age or older. This pharmaceutical benefit is not PBS-subsidised for this condition in a patient who is currently receiving one of the strong CYP3A4 inducers outlined in the Product Information. The authority application must be in writing and must include: (1) a completed authority prescription; and (2) a completed Cystic Fibrosis Authority Application Supporting Information Form; and (3) details of the pathology report substantiating the patient being homozygous for the F508del mutation on the CFTR gene - quote each of the: (i) name of the pathology report provider, (ii) date of pathology report, (iii) unique identifying number/code that links the pathology result to the individual patient; and (4) current CYP3A4 inhibitors, CYP3A4 inducers and IV antibiotics.	Compliance with Written Authority Required procedures

[18] Schedule 3, entry for Ravulizumab

insert in numerical order after existing text:

	C14744		Atypical haemolytic uraemic syndrome (aHUS) Switch from PBS-subsidised eculizumab (all phases) - loading dose Patient must have previously received PBS-subsidised eculizumab under the 'Initial treatment' restriction for this condition; OR Patient must have previously received PBS-subsidised eculizumab under the 'Continuing treatment' restriction for this condition; OR Patient must have previously received PBS-subsidised eculizumab under the 'Extended continuing treatment' restriction for this condition; OR Patient must have previously received PBS-subsidised eculizumab under the 'Recommencement	Compliance with Written Authority Required procedures

			of treatment' restriction for this condition; OR Patient must have previously received PBS-subsidised eculizumab under the 'Continuing recommencement of treatment' restriction for this condition; AND Patient must have/had ADAMTS-13 activity of greater than or equal to 10% on a blood sample; AND Patient must not receive more than 2 weeks of treatment under this restriction. Must be treated by a prescriber who is either: (i) a haematologist, (ii) a nephrologist; OR Must be treated by a medical practitioner who has consulted at least one of the above mentioned specialist types, with agreement reached that the patient should be treated with this pharmaceutical benefit on this occasion; AND Patient must be undergoing treatment with one C5 inhibitor therapy only at any given time. This drug is not PBS-subsidised if it is prescribed to an in-patient in a public hospital setting. The application must indicate the most recent treatment phase that the patient is switching from. For patients who are switching C5 inhibitors, the next application should be sought under the next relevant treatment phase. Serial haematological results (every 3 months while the patient is receiving treatment) must be provided with every subsequent application for treatment. The authority application must be in writing and must include all of the following: (1) A completed authority prescription form(s); (2) A completed authority application form relevant to the indication and treatment phase (the latest version is located on the website specified in the Administrative Advice); (3) A measurement of body weight at the time of application; (4) Results of genetic testing, if not previously submitted.	
	C14746		Atypical haemolytic uraemic syndrome (aHUS) Transitioning from non-PBS to PBS-subsidised treatment - Grandfather arrangements Patient must have previously received non-PBS-subsidised therapy with this drug for this condition; AND Patient must have met all other PBS eligibility criteria that a non-'Grandfather' patient would ordinarily be required to meet, meaning that at the time non-PBS supply was commenced, the patient: (i) had active and progressing thrombotic microangiopathy (TMA) caused by aHUS; (ii) had ADAMTS-13 activity of greater than or equal to 10% on a blood sample not confounded by any plasma exchange or infusion; (iii) had a confirmed negative STEC (Shiga toxin-producing E.Coli) result if the patient has had diarrhoea in the preceding 14 days of commencing ravulizumab treatment; (iv) had clinical features of active organ damage or impairment; AND Patient must have demonstrated ongoing treatment response with ravulizumab for this condition if received at least 26 weeks of initial non-PBS-subsidised therapy; AND Patient must not have experienced treatment failure with ravulizumab for this condition if they have received at least 26 weeks of initial non-PBS-subsidised therapy. Must be treated by a prescriber who is either: (i) a haematologist, (ii) a nephrologist; OR Must be treated by a medical practitioner who has consulted at least one of the above mentioned specialist types, with agreement reached that the patient should be treated with this pharmaceutical benefit on this occasion; AND Patient must be undergoing treatment with one C5 inhibitor therapy only at any given time.	Compliance with Written Authority Required procedures

			This drug is not PBS-subsidised if it is prescribed to an in-patient in a public hospital setting. Evidence of active and progressing TMA is defined by the following: (1) A platelet count of less than 150x10^9/L; and evidence of at least two of the following: (i) presence of schistocytes on blood film; (ii) low or absent haptoglobin; (iii) lactate dehydrogenase (LDH) above normal range; or (2) In recipients of a kidney transplant for end-stage kidney disease due to aHUS, a kidney biopsy confirming TMA; and (3) Evidence of at least one of the following clinical features of active TMA-related organ damage or impairment is defined as below: (a) kidney impairment as demonstrated by one or more of the following: (i) a decline in estimated Glomerular Filtration Rate (eGFR) of greater than 20% in a patient who has pre-existing kidney impairment; (ii) a serum creatinine (sCr) of greater than the upper limit of normal (ULN) in a patient who has no history of pre-existing kidney impairment; (iii) a sCr of greater than the age-appropriate ULN in paediatric patients; (iv) a renal biopsy consistent with aHUS; (b) onset of TMA-related neurological impairment; (c) onset of TMA-related cardiac impairment; (d) onset of TMA-related gastrointestinal impairment; (e) onset of TMA-related pulmonary impairment. Claims of non-renal TMA-related organ damage should be made at the point of application for initial PBS-subsidised ravulizumab (where possible), and should be supported by objective clinical measures. The prescriber's cover letter should establish that the observed organ damage is directly linked to active and progressing TMA, particularly when indirect causes such as severe thrombocytopenia, hypertension and acute renal failure are present at the time of the initial organ impairment. Serial haematological results (every 3 months while the patient is receiving treatment) must be provided with every subsequent application for treatment. The authority application must be in writing and must include all of the following: (1) A completed authority prescription form(s); (2) A completed authority application form relevant to the indication and treatment phase (the latest version is located on the website specified in the Administrative Advice); (3) A detailed cover letter from the prescriber; (4) A measurement of body weight at the time of application; (5) The result of ADAMTS-13 activity on a blood sample taken prior to plasma exchange or infusion; the date and time that the sample for the ADAMTS-13 assay was collected, and the dates and times of any plasma exchanges or infusions that were undertaken in the two weeks prior to collection of the ADAMTS-13 assay; (6) A confirmed negative STEC result if the patient has had diarrhoea in the preceding 14 days of initiating treatment with non-PBS-subsidised ravulizumab; (7) Evidence of active and progressing TMA, including pathology results where relevant. Evidence of the onset of TMA-related neurological, cardiac, gastrointestinal or pulmonary impairment requires a supporting statement with clinical evidence in patient records. All tests must have been	

			performed within 4 weeks of commencement of non-PBS-subsidised ravulizumab; (8) For patients who have received at least 26 weeks of ravulizumab treatment, a recent measurement of eGFR, platelets and two of either LDH, haptoglobin or schistocytes of no more than 1 week old at the time of application.	
	C14747		Atypical haemolytic uraemic syndrome (aHUS) Extended continuing treatment Patient must have received PBS-subsidised ravulizumab under the continuing treatment phase for this condition; OR Patient must have received PBS-subsidised ravulizumab under the switch from eculizumab in the continuing treatment phase for this condition; OR Patient must have received PBS-subsidised ravulizumab under the switch from eculizumab in the extended continuing treatment phase for this condition; AND Patient must have demonstrated ongoing treatment response with PBS-subsidised ravulizumab for this condition; AND Patient must not have experienced treatment failure with ravulizumab for this condition in the most recent treatment phase; AND Patient must have a TMA-related cardiomyopathy as evidenced by left ventricular ejection fraction < 40% on current objective measurement; OR Patient must have severe TMA-related neurological impairment; OR Patient must have severe TMA-related gastrointestinal impairment; OR Patient must have severe TMA-related pulmonary impairment on current objective measurement; OR Patient must have grade 4 or 5 chronic kidney disease (eGFR of less than 30 mL/min); OR Patient must have a high risk of aHUS recurrence in the short term in the absence of continued treatment with ravulizumab; AND Patient must not receive more than 24 weeks of treatment with ravulizumab per continuing treatment course authorised under this restriction. Must be treated by a prescriber who is either: (i) a haematologist, (ii) a nephrologist; OR Must be treated by a medical practitioner who has consulted at least one of the above mentioned specialist types, with agreement reached that the patient should be treated with this pharmaceutical benefit on this occasion; AND Patient must be undergoing treatment with one C5 inhibitor therapy only at any given time. This drug is not PBS-subsidised if it is prescribed to an in-patient in a public hospital setting. A treatment response is defined as: (1) Normalisation of haematology as demonstrated by at least 2 of the following: (i) platelet count, (ii) haptoglobin, (iii) lactate dehydrogenase (LDH); and (2) One of the following: a) an increase in eGFR of > 25% from baseline, where the baseline is the eGFR measurement immediately prior to commencing treatment with a C5 inhibitor; or b) an eGFR within +/- 25% from baseline; or c) an avoidance of dialysis-dependence but worsening of kidney function with a reduction in eGFR 25% from baseline. PBS-subsidised treatment with ravulizumab will not be permitted if a patient has experienced	Compliance with Written Authority Required procedures

			treatment failure with ravulizumab in the most recent treatment phase prior to the treatment phase where this application is sought. A treatment failure is defined as a patient who is: (1) Dialysis-dependent at the time of application and has failed to demonstrate significant resolution of extra-renal complications if originally presented; or (2) On dialysis and has been on dialysis for 4 months of the previous 6 months while receiving a PBS-subsidised C5 inhibitor, and has failed to demonstrate significant resolution of extra-renal complications if originally presented. The authority application must include the following measures of response to the prior course of treatment, including serial haematological results (every 3 months while the patient is receiving treatment). The authority application must be in writing and must include all of the following: (1) A completed authority prescription form(s); (2) A completed authority application form relevant to the indication and treatment phase (the latest version is located on the website specified in the Administrative Advice); (3) A measurement of body weight at the time of application; (4) Results of genetic testing, if not previously submitted; (5) A family history of aHUS, if applicable; (6) A history of multiple episodes of aHUS before commencing ravulizumab treatment, if applicable; (7) A history of kidney transplant, if applicable (especially if required due to aHUS); (8) An inclusion of the individual consequences of recurrent disease; (9) A supporting statement with clinical evidence of severe TMA-related cardiomyopathy (including current LVEF result), neurological impairment, gastrointestinal impairment or pulmonary impairment; (10) Evidence that the patient has had a treatment response including haematological results of no more than 4 weeks old at the time of application (platelet count, haptoglobin and LDH); and an eGFR level of no more than 4 weeks old at the time of application; (11) Evidence that the patient has not experienced treatment failure, including a supporting statement with clinical evidence that the patient does not require dialysis, unless the indication for continuing ravulizumab is severe extra-renal complications that have significantly improved; (12) If the indication for continuing ravulizumab is severe extra-renal complications, then a supporting statement with clinical evidence that any initial extra-renal complications of TMA have significantly improved is required. This assessment must be submitted no later than 4 weeks from the cessation of the prior treatment. Where a response assessment is not undertaken and submitted within these timeframes, the patient will be deemed to have failed to respond to treatment with ravulizumab.	
	C14748		Atypical haemolytic uraemic syndrome (aHUS) Balance of Supply - maintenance doses Patient must have received PBS-subsidised loading dose of ravulizumab for this condition for this current treatment phase; AND Patient must have/had ADAMTS-13 activity of greater than or equal to 10% on a blood sample; AND	Compliance with Written Authority Required procedures

			Patient must have received insufficient therapy to complete the maximum allowable treatment under their specified treatment phase; AND The treatment must provide no more than the balance of up to 24 weeks treatment available under the relevant treatment phase. Must be treated by a prescriber who is either: (i) a haematologist, (ii) a nephrologist; OR Must be treated by a medical practitioner who has consulted at least one of the above mentioned specialist types, with agreement reached that the patient should be treated with this pharmaceutical benefit on this occasion; AND Patient must be undergoing treatment with one C5 inhibitor therapy only at any given time. This drug is not PBS-subsidised if it is prescribed to an in-patient in a public hospital setting. ADAMTS-13 activity result must have been submitted to Services Australia. In the case that a sample for ADAMTS-13 activity taken prior to plasma exchange or infusion was not available at the time of application for Initial treatment, ADAMTS-13 activity must have been measured 7-10 days following the last plasma exchange or infusion and must have been submitted to Services Australia within 13 days of commencement of ravulizumab. The date and time that the sample for the ADAMTS-13 assay was collected, and the dates and times of the last, if any, plasma exchange or infusion that was undertaken in the 2 weeks prior to collection of the ADAMTS-13 assay must also have been provided to Services Australia. Serial haematological results (every 3 months while the patient is receiving treatment) must be provided with every subsequent application for treatment.	
	C14749		Atypical haemolytic uraemic syndrome (aHUS) Continuing treatment Patient must have received PBS-subsidised ravulizumab under the initial treatment phase for this condition; OR Patient must have received PBS-subsidised ravulizumab under the switch from eculizumab in the continuing treatment phase for this condition; OR Patient must have received PBS-subsidised ravulizumab under the grandfather restriction for this condition; AND Patient must have demonstrated ongoing treatment response with PBS-subsidised ravulizumab for this condition; AND Patient must not have experienced treatment failure with ravulizumab for this condition in the most recent treatment phase; AND Patient must not receive more than 72 weeks of ravulizumab treatment in total under this restriction; OR Patient must not receive more than 104 weeks supply of a C5 inhibitor under the initial and continuing treatment restrictions if they had switched C5 inhibitors during the course of initial and continuing treatment; AND Patient must not receive more than 24 weeks of treatment with ravulizumab per continuing treatment course authorised under this restriction. Must be treated by a prescriber who is either: (i) a haematologist, (ii) a nephrologist; OR Must be treated by a medical practitioner who has consulted at least one of the above mentioned specialist types, with agreement reached that the patient should be treated with this pharmaceutical benefit on this occasion; AND	Compliance with Written Authority Required procedures

			Patient must be undergoing treatment with one C5 inhibitor therapy only at any given time. This drug is not PBS-subsidised if it is prescribed to an in-patient in a public hospital setting. A treatment response is defined as: (1) Normalisation of haematology as demonstrated by at least 2 of the following: (i) platelet count, (ii) haptoglobin, (iii) lactate dehydrogenase (LDH); and (2) One of the following: a) an increase in eGFR of > 25% from baseline, where the baseline is the eGFR measurement immediately prior to commencing treatment with a C5 inhibitor; or b) an eGFR within +/- 25% from baseline; or c) an avoidance of dialysis-dependence but worsening of kidney function with a reduction in eGFR 25% from baseline. PBS-subsidised treatment with ravulizumab will not be permitted if a patient has experienced treatment failure with ravulizumab in the most recent treatment phase prior to the treatment phase where this application is sought. A treatment failure is defined as a patient who is: (1) Dialysis-dependent at the time of application and has failed to demonstrate significant resolution of extra-renal complications if originally presented; or (2) On dialysis and has been on dialysis for 4 months of the previous 6 months while receiving a PBS-subsidised C5 inhibitor, and has failed to demonstrate significant resolution of extra-renal complications if originally presented. The authority application must include the following measures of response to the prior course of treatment, including serial haematological results (every 3 months while the patient is receiving treatment). The authority application must be in writing and must include all of the following: (1) A completed authority prescription form(s); (2) A completed authority application form relevant to the indication and treatment phase (the latest version is located on the website specified in the Administrative Advice); (3) A measurement of body weight at the time of application; (4) Results of genetic testing, if not previously submitted; (5) A family history of aHUS, if applicable; (6) A history of kidney transplant if applicable (especially if required due to aHUS); (7) An inclusion of the individual consequences of recurrent disease, if applicable; (8) Evidence that the patient has had a treatment response including haematological results of no more than 1 week old at the time of application (platelet count, haptoglobin and LDH); and an eGFR level of no more than 1 week old at the time of application; (9) Evidence that the patient has not experienced treatment failure, including a supporting statement with clinical evidence that the patient does not require dialysis, unless the indication for continuing ravulizumab is severe extra-renal complications that have significantly improved; (10) If the indication for continuing ravulizumab is severe extra-renal complications, then a supporting statement with clinical evidence that any initial extra-renal complications of TMA have significantly improved is required. This assessment must be submitted no later than 4 weeks from the cessation of the prior treatment. Where a response assessment is not undertaken and submitted within these timeframes, the patient will be deemed to have failed to respond to treatment with ravulizumab.	

	C14780		Atypical haemolytic uraemic syndrome (aHUS) Initial treatment - Initial (new patient) loading dose Patient must have active and progressing thrombotic microangiopathy (TMA) caused by aHUS; AND Patient must have ADAMTS-13 activity of greater than or equal to 10% on a blood sample taken prior to plasma exchange or infusion; or, if ADAMTS-13 activity was not collected prior to plasma exchange or infusion, patient must have platelet counts of greater than 30x10^9/L and a serum creatinine of greater than 150 mol/L; AND Patient must have a confirmed negative STEC (Shiga toxin-producing E.Coli) result if the patient has had diarrhoea in the preceding 14 days; AND Patient must have clinical features of active organ damage or impairment; AND Patient must not receive more than 2 weeks of treatment under this restriction. Must be treated by a prescriber who is either: (i) a haematologist, (ii) a nephrologist; OR Must be treated by a medical practitioner who has consulted at least one of the above mentioned specialist types, with agreement reached that the patient should be treated with this pharmaceutical benefit on this occasion; AND Patient must be undergoing treatment with one C5 inhibitor therapy only at any given time. This drug is not PBS-subsidised if it is prescribed to an in-patient in a public hospital setting. Evidence of active and progressing TMA is defined by the following: (1) A platelet count of less than 150x10^9/L; and evidence of at least two of the following: (i) presence of schistocytes on blood film; (ii) low or absent haptoglobin; (iii) lactate dehydrogenase (LDH) above normal range; or (2) In recipients of a kidney transplant for end-stage kidney disease due to aHUS, a kidney biopsy confirming TMA; and (3) Evidence of at least one of the following clinical features of active TMA-related organ damage or impairment is defined as below: (a) kidney impairment as demonstrated by one or more of the following: (i) a decline in estimated Glomerular Filtration Rate (eGFR) of greater than 20% in a patient who has pre-existing kidney impairment; (ii) a serum creatinine (sCr) of greater than the upper limit of normal (ULN) in a patient who has no history of pre-existing kidney impairment; (iii) a sCr of greater than the age-appropriate ULN in paediatric patients; (iv) a renal biopsy consistent with aHUS; (b) onset of TMA-related neurological impairment; (c) onset of TMA-related cardiac impairment; (d) onset of TMA-related gastrointestinal impairment; (e) onset of TMA-related pulmonary impairment. Claims of non-renal TMA-related organ damage should be made at the point of application for initial PBS-subsidised ravulizumab (where possible), and should be supported by objective clinical measures. The prescriber's cover letter should establish that the observed organ damage is directly linked to active and progressing TMA, particularly when indirect causes such as severe thrombocytopenia, hypertension and acute renal failure are present at the time of the initial organ impairment.	Compliance with Written Authority Required procedures

			Serial haematological results (every 3 months while the patient is receiving treatment) must be provided with every subsequent application for treatment. The authority application must be in writing and must include all of the following: (1) A completed authority prescription form(s); (2) A completed authority application form relevant to the indication and treatment phase (the latest version is located on the website specified in the Administrative Advice); (3) A detailed cover letter from the prescriber; (4) A measurement of body weight at the time of application; (5) The result of ADAMTS-13 activity on a blood sample taken prior to plasma exchange or infusion; the date and time that the sample for the ADAMTS-13 assay was collected, and the dates and times of any plasma exchanges or infusions that were undertaken in the 2 weeks prior to collection of the ADAMTS-13 assay; (6) In the case that a sample for ADAMTS-13 assay was not collected prior to plasma exchange or infusion, measurement of ADAMTS-13 activity must be taken 7-10 days following the last plasma exchange or infusion. The ADAMTS-13 result must be submitted to Services Australia within 13 days of commencement of ravulizumab treatment in order for the patient to be considered as eligible for further PBS-subsidised C5 inhibitor treatment, under Initial balance of supply; (7) A confirmed negative STEC result if the patient has had diarrhoea in the preceding 14 days; (8) Evidence of active and progressing TMA, including pathology results where relevant. Evidence of the onset of TMA-related neurological, cardiac, gastrointestinal or pulmonary impairment requires a supporting statement with clinical evidence in patient records. All tests must have been performed within 4 weeks of application; (9) For all patients, a recent measurement of eGFR, platelets and two of either LDH, haptoglobin or schistocytes of no more than 1 week old at the time of application. Two authority prescription forms will be required to cover for the 26 weeks of initial therapy with ravulizumab, one for the loading dose and one for the 24 week balance which can be sought under the Balance of Supply.	
	C14791		Atypical haemolytic uraemic syndrome (aHUS) Recommencement of treatment Patient must have demonstrated treatment response to previous treatment with a PBS-subsidised C5 inhibitor for this condition; AND Patient must not have experienced treatment failure with ravulizumab for this condition in the most recent treatment phase; AND Patient must have the following clinical conditions prior to recommencing C5 inhibitor treatment: (i) either significant haemolysis as measured by low/absent haptoglobin; or presence of schistocytes on the blood film; or lactate dehydrogenase (LDH) above normal; AND (ii) either platelet consumption as measured by either 25% decline from patient baseline or thrombocytopenia (platelet count <150 x 10^9/L); OR (iii) TMA-related organ impairment including on recent biopsy. Must be treated by a prescriber who is either: (i) a haematologist, (ii) a nephrologist; OR Must be treated by a medical practitioner who has consulted at least one of the above mentioned specialist types, with agreement reached that the patient should be treated with this pharmaceutical benefit on this occasion; AND	Compliance with Written Authority Required procedures

			Patient must be undergoing treatment with one C5 inhibitor therapy only at any given time. This drug is not PBS-subsidised if it is prescribed to an in-patient in a public hospital setting. A treatment response is defined as: (1) Normalisation of haematology as demonstrated by at least 2 of the following: (i) platelet count, (ii) haptoglobin, (iii) lactate dehydrogenase (LDH); and (2) One of the following: a) an increase in eGFR of > 25% from baseline, where the baseline is the eGFR measurement immediately prior to commencing treatment with a C5 inhibitor; or b) an eGFR within +/- 25% from baseline; or c) an avoidance of dialysis-dependence but worsening of kidney function with a reduction in eGFR 25% from baseline. PBS-subsidised treatment with ravulizumab will not be permitted if a patient has experienced treatment failure with ravulizumab in the most recent treatment phase prior to the treatment phase where this application is sought. A treatment failure is defined as a patient who is: (1) Dialysis-dependent at the time of application and has failed to demonstrate significant resolution of extra-renal complications if originally presented; or (2) On dialysis and has been on dialysis for 4 months of the previous 6 months while receiving a PBS-subsidised C5 inhibitor, and has failed to demonstrate significant resolution of extra-renal complications if originally presented. The authority application must include the following measures of response to the prior course of treatment, including serial haematological results (every 3 months while the patient is receiving treatment). The authority application must be in writing and must include all of the following: (1) A completed authority prescription form(s); (2) A completed authority application form relevant to the indication and treatment phase (the latest version is located on the website specified in the Administrative Advice); (3) A measurement of body weight at the time of application; (4) Results of genetic testing, if not previously submitted; (5) A family history of aHUS if applicable; (6) A history of multiple episodes of aHUS following the treatment break, if applicable; (7) A history of kidney transplant if applicable (especially if required due to aHUS); (8) An inclusion of the individual consequences of recurrent disease; (9) A supporting statement with clinical evidence of TMA-related organ damage including current (within one week of application) haematological results (platelet count, haptoglobin and LDH), eGFR level, and, if applicable, on recent biopsy; (10) Evidence that the patient has had a treatment response to their previous treatment with a C5 inhibitor; (11) Evidence that the patient has not experienced treatment failure, including a supporting statement with clinical evidence that the patient does not require dialysis, unless the indication for continuing ravulizumab is severe extra-renal complications that have significantly improved; (12) If the indication for continuing ravulizumab is severe extra-renal complications, then a supporting statement with clinical evidence that any initial extra-renal complications of TMA have significantly improved is required.	

			Two authority prescription forms will be required to cover for the 26 weeks of recommencement therapy with ravulizumab, one for the loading dose and one for the 24 week balance which can be sought under the Balance of Supply.	
	C14797		Atypical haemolytic uraemic syndrome (aHUS) Continuing recommencement of treatment Patient must have received PBS-subsidised ravulizumab under the 'Recommencement of treatment' restriction for this condition; OR Patient must have received PBS-subsidised ravulizumab under the switch from eculizumab 'Recommencement treatment' restriction for this condition; OR Patient must have received PBS-subsidised ravulizumab under the switch from eculizumab 'Continuing recommencement treatment' restriction for this condition; AND Patient must have demonstrated ongoing treatment response to 'Recommencement of treatment' with a C5 inhibitor for this condition; AND Patient must not have experienced treatment failure with ravulizumab for this condition in the most recent treatment phase; AND Patient must not receive more than 24 weeks of treatment with ravulizumab per continuing treatment course authorised under this restriction. Must be treated by a prescriber who is either: (i) a haematologist, (ii) a nephrologist; OR Must be treated by a medical practitioner who has consulted at least one of the above mentioned specialist types, with agreement reached that the patient should be treated with this pharmaceutical benefit on this occasion; AND Patient must be undergoing treatment with one C5 inhibitor therapy only at any given time. This drug is not PBS-subsidised if it is prescribed to an in-patient in a public hospital setting. A treatment response is defined as: (1) Normalisation of haematology as demonstrated by at least 2 of the following: (i) platelet count, (ii) haptoglobin, (iii) lactate dehydrogenase (LDH); and (2) One of the following: a) an increase in eGFR of > 25% from baseline, where the baseline is the eGFR measurement immediately prior to commencing treatment with a C5 inhibitor; or b) an eGFR within +/- 25% from baseline; or c) an avoidance of dialysis-dependence but worsening of kidney function with a reduction in eGFR 25% from baseline. PBS-subsidised treatment with ravulizumab will not be permitted if a patient has experienced treatment failure with ravulizumab in the most recent treatment phase prior to the treatment phase where this application is sought. A treatment failure is defined as a patient who is: (1) Dialysis-dependent at the time of application and has failed to demonstrate significant resolution of extra-renal complications if originally presented; or (2) On dialysis and has been on dialysis for 4 months of the previous 6 months while receiving a PBS-subsidised C5 inhibitor, and has failed to demonstrate significant resolution of extra-renal complications if originally presented. The authority application must include the following measures of response to the prior course of treatment, including serial haematological results (every 3 months while the patient is receiving	Compliance with Written Authority Required procedures

			treatment). The authority application must be in writing and must include all of the following: (1) A completed authority prescription form(s); (2) A completed authority application form relevant to the indication and treatment phase (the latest version is located on the website specified in the Administrative Advice); (3) A measurement of body weight at the time of application; (4) Results of genetic testing, if not previously submitted; (5) A family history of aHUS, if applicable; (6) A history of multiple episodes of aHUS before recommencing ravulizumab treatment, if applicable; (7) A history of kidney transplant if applicable (especially if required due to aHUS); (8) An inclusion of the individual consequences of recurrent disease, if applicable; (9) Evidence that the patient has had a treatment response including haematological results of no more than 1 week old at the time of application (platelet count, haptoglobin and LDH); and an eGFR level of no more than 1 week old at the time of application; (10) Evidence that the patient has not experienced treatment failure, including a supporting statement with clinical evidence that the patient does not require dialysis, unless the indication for continuing ravulizumab is severe extra-renal complications that have significantly improved; (11) If the indication for continuing ravulizumab is severe extra-renal complications, then a supporting statement with clinical evidence that any initial extra-renal complications of TMA have significantly improved is required. This assessment must be submitted no later than 4 weeks from the cessation of the prior treatment. Where a response assessment is not undertaken and submitted within these timeframes, the patient will be deemed to have failed to respond to treatment with ravulizumab	

[19] Schedule 3, entry for Ustekinumab

insert in numerical order after existing text:

	C14758		Complex refractory Fistulising Crohn disease Initial treatment - Initial 2 (change or recommencement of treatment after a break in biological medicine of less than 5 years) Patient must have received prior PBS-subsidised treatment with a biological medicine for this condition in this treatment cycle; AND Patient must not have failed PBS-subsidised therapy with this drug for this condition more than once in the current treatment cycle. Must be treated by a gastroenterologist (code 87); OR Must be treated by a consultant physician [internal medicine specialising in gastroenterology (code 81)]; OR Must be treated by a consultant physician [general medicine specialising in gastroenterology (code 82)]. To demonstrate a response to treatment the application must be accompanied with the assessment of response, conducted between 8 and 16 weeks of therapy and no later than 4 weeks from cessation of the most recent course of biological medicine. It is recommended that an	Compliance with Written Authority Required procedures

			application for the continuing treatment be submitted no later than 4 weeks from the date of completion of the most recent course of treatment. This is to ensure treatment continuity for those who meet the continuing restriction. Where a response assessment is not conducted within the required timeframe, the patient will be deemed to have failed to respond to treatment with this drug, unless the patient has experienced a serious adverse reaction of a severity resulting in the necessity for permanent withdrawal of treatment. Applications for authorisation must be made in writing and must include: (1) two completed authority prescription forms; and (2) a completed authority application form relevant to the indication and treatment phase (the latest version is located on the website specified in the Administrative Advice) which includes the following: (i) a completed current Fistula Assessment Form including the date of assessment of the patient's condition; and (ii) details of prior biological medicine treatment including details of date and duration of treatment. Two completed authority prescriptions should be submitted with every initial application for this drug. One prescription should be written under S100 (Highly Specialised Drugs) for a weight-based loading dose, containing a quantity of up to 4 vials of 130 mg and no repeats. The second prescription should be written under S85 (General) for 1 vial or pre-filled syringe of 90 mg and no repeats. The most recent fistula assessment must be no more than 4 weeks old at the time of application. A maximum quantity of a weight-based loading dose is up to 4 vials with no repeats and the subsequent first dose of 90 mg with no repeats provide for an initial 16-week course of this drug will be authorised Where fewer than 6 vials in total are requested at the time of the application, authority approvals for a sufficient number of vials based on the patient's weight to complete dosing at weeks 0 and 8 may be requested by telephone through the balance of supply restriction. Under no circumstances will telephone approvals be granted for initial authority applications, or for treatment that would otherwise extend the initial treatment period.	
	C14787		Complex refractory Fistulising Crohn disease Initial treatment - Initial 1 (new patient or recommencement of treatment after a break in biological medicine of more than 5 years) Patient must have confirmed Crohn disease, defined by standard clinical, endoscopic and/or imaging features, including histological evidence, with the diagnosis confirmed by a gastroenterologist or a consultant physician; AND Patient must have an externally draining enterocutaneous or rectovaginal fistula. Must be treated by a gastroenterologist (code 87); OR Must be treated by a consultant physician [internal medicine specialising in gastroenterology (code 81)]; OR Must be treated by a consultant physician [general medicine specialising in gastroenterology (code 82)]. Applications for authorisation must be made in writing and must include: (1) two completed authority prescription forms; and	Compliance with Written Authority Required procedures

			(2) a completed authority application form relevant to the indication and treatment phase (the latest version is located on the website specified in the Administrative Advice) which includes a completed current Fistula Assessment Form including the date of assessment of the patient's condition of no more than 4 weeks old at the time of application. Two completed authority prescriptions should be submitted with every initial application for this drug. One prescription should be written under S100 (Highly Specialised Drugs) for a weight-based loading dose, containing a quantity of up to 4 vials of 130 mg and no repeats. The second prescription should be written under S85 (General) for 1 vial or pre-filled syringe of 90 mg and no repeats. An assessment of a patient's response to this initial course of treatment must be conducted between 8 and 16 weeks of therapy. Where a response assessment is not conducted within the required timeframe, the patient will be deemed to have failed to respond to treatment with this drug, unless the patient has experienced a serious adverse reaction of a severity resulting in the necessity for permanent withdrawal of treatment. A maximum quantity of a weight-based loading dose is up to 4 vials with no repeats and the subsequent first dose of 90 mg with no repeats provide for an initial 16-week course of this drug will be authorised Where fewer than 6 vials in total are requested at the time of the application, authority approvals for a sufficient number of vials based on the patient's weight to complete dosing at weeks 0 and 8 may be requested by telephone through the balance of supply restriction. Under no circumstances will telephone approvals be granted for initial authority applications, or for treatment that would otherwise extend the initial treatment period.	
	C14801		Complex refractory Fistulising Crohn disease Initial 1 (new patient or recommencement of treatment after a break in biological medicine of more than 5 years), Initial 2 (change or recommencement of treatment after a break in biological medicine of less than 5 years) - balance of supply Patient must have received insufficient therapy with this drug for this condition under the Initial 1 (new patient or patient recommencing treatment after a break of 5 years or more) restriction to complete 16 weeks treatment; OR Patient must have received insufficient therapy with this drug for this condition under the Initial 2 (change or recommencement of treatment after a break of less than 5 years) restriction to complete 16 weeks treatment; AND The treatment must provide no more than the balance of up to 16 weeks treatment available under the above restrictions. Must be treated by a gastroenterologist (code 87); OR Must be treated by a consultant physician [internal medicine specialising in gastroenterology (code 81)]; OR Must be treated by a consultant physician [general medicine specialising in gastroenterology (code 82)]	Compliance with Authority Required procedures