Japan), Speakers bureau: Tanabe Mitsubishi, Bristol-Meyers, Toshihiro Matsui Speakers bureau: Takeda, Chugai, Eisai, Pfizer, Astellas, Eli Lilly, Ono, AbbVie, Mitsubishi Tanabe, AsahiKASEI, UCB, Teijin, Susumu Nishiyama Paid instructor for: Kissei Pharmaceutical Co., Ltd, Takeda Pharmaceutical Company, Mitsubishi Tanabe Pharma, Astellas Pharma, Eisai Pharmaceutical company, AbbVie Inc., TEIJIN Pharma, Asahi Kasei Corporation, Bristol-Myers Squibb Company, Chugai Pharmaceutical Co., Ltd., Santen Pharmaceutical Co., Ltd., AYUMI Pharmaceutical Corporation, Speakers bureau: Kissei Pharmaceutical Co., Ltd, DAIICHI SANKYO COMPANY, LIMITED, Shiro Ohshima Grant/research support from: AbbVie, Eisai, Asahikasei, Speakers bureau: AbbVie, Eisai, Bristol-Meyers, Novartis, Astellas, Nippon-Kayaku, Pfizer, UCB, Ayumi, Daiichi-Sankyo, Takeda, Tanabe-Matsubishi, Chugai, Hiroaki Umebayashi Speakers bureau: AbbVie, Eisai, Novartis, Ono, Chugai, Takahiro Yasumi Shareholder of: Takeda, Speakers bureau: AbbVie, Novartis, CSL Behring, Masaaki Mori Grant/research support from: Tokyo Medical and Dental University (TMDU) received unrestricted research grants for Department of Lifetime Clinical Immunology from AbbVie GK, Ayumi Pharmaceutical Corporation, Chugai Pharmaceutical Co., Ltd., CSL Behring K.K., Japan Blood Products Organization, Mitsubishi Tanabe Pharma Corporation, Nippon Kayaku Co., Ltd., Ono Pharmaceutical Co., Ltd., Towa Pharmaceutical Co., Ltd., UCB Japan Co. Ltd.

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AB1027 DEVELOPMENT OF A NOVEL DIAGNOSTIC METHOD FOR ATYPICAL AND TREATMENT-REFRACTORY KAWASAKI DISEASE USING NEWLY IDENTIFIED PROTEINS AS BIOMARKERS

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Background: Kawasaki disease (KD) is a representative acute inflammatory disease of the children mainly under 4 years old. There were more than 15,000 onset cases per year in Japan. KD without proper treatment can lead to cardiovascular complications such as coronary artery lesions (CALs). We are still missing about an etiology and the onset mechanism of KD. Therefore, the diagnosis of KD depends on its clinical features. There are also no specific biomarkers for therapy effectiveness in clinical settings. By using proteomic analysis, we found that the serum expression of lipopolysaccharide binding protein (LBP) and leucine rich α 2 - glycoprotein 1 (LRG1) at the acute phase of KD were significantly high, compared with that of healthy control or patients with autoimmune disease and these two proteins could diagnose KD efficiently. We also found the usefulness of LBP and LRG1 values as KD-related biomarkers for therapy effectiveness.

Objectives: To develop a novel objective and quick diagnostic method using LBP and LRG1 for "atypical case" that cause a CALs without presenting symptoms of typical KD, or "treatment-refractory case" that is none-effective for immunoglobulin therapy.

Methods: The research strategy is composed of clinical phenotype sub-grouping of KD patients using clustering analysis based on clinical features and biomarkers such as LRG1 and LBP. This study contains more than 350 KD patients' data sets including clinical characteristics and laboratory parameters including those of Biomarkers.

Results: According to these results of study, we can objectively diagnose KD without depending on only clinical manifestations.

Conclusion: These two biomarkers may be added in a new KD diagnostic criteria, resulting in quality enhancement of KD treatment and may have the potential of proper clinical examination for KD without consequence of CALs.

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AB1028 MYOCARDITIS IN PEDIATRIC LUPUS: A CLINICAL CONUNDRUM

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Background: Myocarditis in pediatric lupus is uncommon but carry a significant morbidity and mortality.

Objectives: To study the clinical profile and outcome of children with lupus myocarditis

Methods: We reviewed our cohort of children with lupus registered between January 1993 to November 2018. Of the 140 lupus patients that were diagnosed during this time, 3 had myocarditis.

Results: Patient 1 was an 11-year-old girl who presented with fever, rash and generalized body swelling for 3 months. Physical examinations revealed heart rate 148/min, respiratory rate 30/min, and blood pressure (BP) 100/80 mmHg. She had pallor, malar rash, hepatomegaly and gallop rhythm. Investigations are detailed in the table. She received intravenous immunoglobulin and initiated on pulse methylprednisolone and followed by pulse intravenous cyclophosphamide. She was discharged on oral prednisolone and subsequently tapered. Patient 2 was a 9-year-old girl presented with fever and rash for 3 months with lethargy for 1 day. She had pallor, malar rash, hepatomegaly. Physical examinations revealed heart rate 132/min, respiratory rate 32/min, feeble peripheral pulses and BP 100/80 mmHg. She received intravenous cyclophosphamide monthly for 6 months along with oral prednisolone in tapering dose. Patient 3 was a 12-year-old girl, who was known to have SLE for 2 months, presented with fever, malar rash, and photosensitivity. She had stopped taking steroids for 2 weeks. On examination, she had heart rate 122/min, respiratory rate 28/min and BP 100/72 mmHg. She had pallor, alopecia, oral ulcers, malar rash, gallop rhythm and tender hepatomegaly. She was given pulse methylprednisolone and furosemide therapy followed by oral prednisolone in gradually tapering dose.

Conclusion: Myocarditis in lupus is uncommon but carries a significant morbidities. The presence of global hypokinesia with a low ejection fraction on two dimensional echocardiography (2DE) is a strong pointer towards the diagnosis. Immunosuppressant drugs with supportive therapy remain the preferred treatment. Early diagnosis and prompt treatment may be very rewarding.

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Table: Laboratory investigations:

Investigation	Case 1	Case 2	Case 3
Haemoglobin (g/L)	94g/L	92 g/L	80 g/L
White cell counts	6.7 10 ⁹ /L	7.2 10 ⁹ /L	6.5 10 ⁹ /L
Lymphocyte count	1.15x 10 ⁹ /L	1.91 X 10 ⁹ /L	1.65X10 ⁹ /L
Platelets	130x10 ⁹ /L	150 x10 ⁹ /L	220 x 10 ⁹ /L
Urine routine Urine protein (mg/m ² /hour)	No RBC, No albumin	No RBC, No albumin	No RBC, No albumin
C3 (Normal 50-150 mg/dL)	27 mg/dl	28 mg/dL	31 mg/dl
C4 (Normal 20-50 mg/dL)	4 mg/dl	3 mg/dL	10 mg/dl
ANA	4+ diffuse	3 + diffuse	3+ diffuse
Anti dsDNA (N: <25 IU/mL)	59	583	112
Antiphospholipid antibodies: a) Lupus anticoagulant b) Anticardiolipin antibody (IgG and IgM) c) Anti B2 Glycoprotein -1 antibody (IgG & IgM)	Positive Negative Negative	Negative Negative Negative	Negative Negative Negative
2Dimensional Echocardiography	Mild pericardial effusion, dilated left ventricle, ejection fraction (EF) 30%	Mild pericardial effusion, EF 25%	Mild pericardial effusion, moderate mitral regurgitation, mild tricuspid regurgitation, global hypokinesia, EF 56%

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