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| Table 1. Results of the 32 Cases, Published During the 2020–2022 Period, Including Thrombotic Thrombocytopenic Purpura (TTP) following COVID-19 Vaccination | | | | | | | | | | |
| Authors and Ref | **Country** | **Old Gender** | **Underlying disease** | **First Episode**  **Relapse** | **Symptoms** | **Vaccine**  **Dose**  **Time after vaccination** | **Biology** | **ADAMTS 13 activity**  **Autoantibody\*** | **Treatments** | **Outcome** |
| Chamarti et al. | USA  (2021) | 80  Male | Hypertension  Diabetes  Hyperlipidemia  Gout  Iron deficiency  Anemia | First | Generalized weakness  Malaise | Pfizer-BioNTech  Second dose  14 days | Hemoglobin, 4.8 g/dl  Platelets, 48\*109/L  Schistocytes, +++  Creatinine, 212.16 µmol/L  LDH, 1118 UI/l  Haptoglobin, <10 mg/dl | <2%  182 U/ml | Plasma Exchange Steroids  Rituximab | Improved |
| Giuffrida et al. | Italy  (2021) | 83  Female | Undifferentiated connective tissue disease  Diabetes | First | Severe anemia  Macro-hematuria  Diffuse petechiae | Pfizer-BioNTech  First dose  7 days | Hemoglobin, 6.1 g/dl  Reticu, 28%  Platelets, 46\*109/L  Schistocytes, 10%  Creatinine, 77.79 µmol/L  LDH, 1905 UI/l  Haptoglobin, <7 mg/dl | <10%  40 U/ml | Plasma Exchange Steroids  Caplacizumab | Death (probably due to a  sudden cardiovascular event) |
| 30  Female | Beta-thalassemia | First | Diffuse petechiae  Intense headache  Fatigue | Pfizer-BioNTech  First dose  18 days | Hemoglobin, 8.9 g/dl  Retic, 29%  Platelets, 11\*109/L  Schistocytes, 5-10%  Creatinine, 79.56 µmol/L  LDH, 900 UI/l  Haptoglobin, <7 mg/dl | <10%  77.6 U/ml | Plasma Exchange Steroids  Caplacizumab | Improved |
| Karabulut et al. | USA (2021) | 48  Male | No | First | Acute-onset, transient right-sided weakness  Slurred speech lasting | Moderna Biotech  First dose  5 days | Hemoglobin, 8.8 g/dl  Platelets, 10\*109/L  Schistocytes, 2-3%  Creatinine, 83.98 µmol/L  LDH, 884 UI/l  Haptoglobin, <10 mg/dl | <3%  6.6 BEU | Plasma Exchange Steroids  Rituximab | Improved |
| Lee et al. | UK (2021) | 50  Female | Hypertension | First | Dysphasia  Acute upper limb numbness | AstraZeneca  First dose  12 days | Hemoglobin, 9.9 g/dl  Retic, 6.9%  Platelets, 33\*109/L  Schistocytes, +  LDH, 359 UI/l | 0%  94.93 U/ml | Plasma Exchange Steroids  Rituximab | Improved |
| Maayan et al. | Israel  (2021) | 40  Female | No | First | Somnolence  Fever  Macroscopic hematuria | Pfizer-BioNTech  Second dose  8 days | Hemoglobin, 9.9 g/dl  Platelets, 12\*109/L  Schistocytes, 6%  Creatinine, 81.35 µmol/L  LDH, 7129 UI/l | 0%  51 U/ml | Plasma Exchange  Steroids  Caplacizumab | Improved |
| 28  Male | Morbid obesity | First | Dysarthria | Pfizer-BioNTech  Second dose  28 days | Hemoglobin, 9.1 g/dl  Platelets, 38\*109/L  Schistocytes, 6%  Creatinine, 132.63 µmol/L  LDH, 3063 UI/l | 0%  113 U/ml | Plasma Exchange Steroids  Caplacizumab Rituximab | Improved |
| 31  Female | TTP | Relapse | Vaginal bleeding  Purpura | Pfizer-BioNTech  First dose  13 days | Hemoglobin, 7.7 g/dl  Platelets, 17\*109/L  Schistocytes, 10%  Creatinine, 106 µmol/L  LDH, 4000 UI/l | 0%  64 U/ml | Plasma Exchange Steroids  Caplacizumab  Rituximab | Continu caplacizumab |
| 30  Male | TTP | Relapse | Purpura | Pfizer-BioNTech  Second dose  8 days | Hemoglobin, 8.3 g/dl  Retic, 8%  Platelets, 14\*109/L  Schistocytes, 14%  Renal function, normal  LDH, 1138 UI/l | 0%  21 U/ml | Plasma Exchange Steroids  Caplacizumab  Rituximab | Improved |
| Osmanodja et al. | Germany  (2021) | 25  Male | No | First | Persisting malaise  Fever  Headache  Word-finding difficulties  Nausea, vomiting  Petechial bleeding  Hematuria | Moderna Biotech  First dose  13 days | Hemoglobin, 7.4 g/dl  Retic, 233.1 109/L  Platelets, 29\*109/L  Schistocytes, 2.1%  Creatinine, 132.6 µmol/L  LDH, 999 UI/l  Haptoglobin, <8 mg/dl | <5%  72.2 U/ml | Plasma Exchange Steroids  Caplacizumab  Rituximab | Continu caplacizumab |
| Pavenski et al. | Canada (2021) | 84  Male | TTP  Prostate cancer Hypertension Diabetes  Gout  Hypercholesterolemia | Relapse | Lethargy  Myalgias  Anorexia | Pfizer-BioNTech  First dose  7 days | Hemoglobin, 7.2 g/dl  Retic, elevated  Platelets, 58\*109/L  Schistocytes, +  Creatinine, 77 µmol/L  LDH, 594 UI/l | <1%  >15 U/ml | Plasma Exchange  Steroids  Rituximab | Improved |
| Sissa et al. | Italy (2021) | 48  Female | TTP | Relapse | Ecchymosis | Pfizer-BioNTech  Second dose  6 days | Hemoglobin, 11.5 g/dl  Platelets, 94\*109/L  Schistocytes, 10%  Renal function, normal  LDH, 637 UI/l | <3%  88 U/ml | Plasma Exchange  Steroids | Improved |
| Waqar et al. | USA  (2021) | 69  Male | Hypertension Chronic kidney disease  HIV  Chronic hepatitis B  Deep  vein thrombosis | First | Severe fatigue  Shortness of breath | Pfizer-BioNTech  Second dose  7 days | Hemoglobin, 9.3 g/dl  Retic, 2.8%  Platelets, 22\*109/L  Schistocytes, ++  Creatinine, 177.68 µmol/L  LDH, 1229 UI/l | 2%  >90 U/ml | Plasma Exchange Steroids  Rituximab | Improved |
| Yucum et al. | USA (2021) | 62  Female | Hypertension  Hyperlipidemia  Hypothyroidism | first | Acute onset of altered mental status | Johnson and Johnson  First dose  37 days | Hemoglobin, 8.2 g/dl  Retic, 8%  Platelets, 11\*109/L  Creatinine, 530 µmol/L  LDH, > 2500 UI/l  ASAT/ALAT, 982 / 231 U/L | <12%  NA | Plasma Exchange Steroids  Hemodialysis | Improved |
| Al Ahmad et al. | Kuwait (2021) | 37  Male | Secondary polycythemia | first | Dizziness, fatigue  Headache  Shortness of breath  Palpitation  Dark urine and petechiae | AstraZeneca-Oxford  First dose  10 days | Hemoglobin, 8.3 g/dl  Retic, 8%  Platelets, 14\*109/L  Schistocytes, 14%  Renal function, normal  LDH, 1138 UI/l | 2.6%  Positive | Plasma Exchange Steroids  Rituximab | Improved |
| De Bruijn et al. | Belgium  (2021) | 38  Female | No | First | Spontaneous  bruising and petechiae | Pfizer-BioNTech  First dose  14 days | Hemoglobin, 10.5 g/dl  Retic, 263 109/L  Platelets, 46\*109/L  Schistocytes, 3%  Creatinine, 83.98 µmol/L  LDH, 631 UI/l | 0%  106.8 BEU | Plasma Exchange  Steroids  Caplacizumab  Rituximab | Improved |
| Alislambouli et al. | USA  (2022) | 61  Male | No | First | Confusion  Fever  Headache  Emesis  Dark urine  Leg ecchymosis | Pfizer-BioNTech  First dose  5 days | Hemoglobin, 6.5 g/dl  Retic, 8%  Platelets, 6\*109/L  Schistocytes, 8%  LDH, 1757 UI/l  Haptoglobin, <8 mg/dl | <3%  NA | Plasma Exchange  Steroids  Rituximab | Improved |
| Deucher et al. | USA  (2022) | 28  Female | TTP | Relapse | Bruising on arms | Pfizer-BioNTech First dose  5 days | Hemoglobin, 10.5 g/dl  Platelets, 84\*109/L  Schistocytes, ++  LDH, 205 UI/l  Haptoglobin, undetectable | <2.5%  Positive | Caplacizumab  Steroids  Rituximab | Improved |
| Innao et al. | Italy  (2022) | 33  Female | Hodgkin Lymphoma  Gray Zone Lymphoma | First | Asthenia  Drowsiness  Headache  Nausea  Abdominal pain  Lower extremity purpura | Pfizer-BioNTech  First dose  9 days | Hemoglobin, 6.8 g/dl  Retic, 896\*109/L  Platelets, 12\*109/L  Schistocytes, 3%  Creatinine, 122 µmol/L  LDH, 1280 UI/l  Haptoglobin, <6 mg/dl | 8%  5 U/ml (not valuable, due to defects in the sample) | Plasma Exchange  Steroids  Caplacizumab | Improved |
| Kirpalani et al. | Japan  (2022) | 14  Female | Anxiety  Iron  Deficiency | First | Fatigue  Headache  Confusion  Bruising | Pfizer-BioNTech  First dose  14 days | Hemoglobin, 6.3 g/dl  Platelets, 10\*109/L  Schistocytes, +  LDH, 626 UI/l  Haptoglobin, <10 mg/dl | <1%  72 U/ml | Plasma Exchange Steroids  Caplacizumab  Rituximab | Improved |
| Ruhe et al. | Germany (2022) | 84  Female | No | First | Partial hemiplegia  Scattered petechiae | Pfizer-BioNTech  First dose  16 days | Hemoglobin, 7.9 g/dl  Platelets, 45\*109/L  Schistocytes, 4.2%  Creatinine, 172.38 µmol/L  Haptoglobin, <10 mg/dl | 1.6%  82.2 U/ml | Plasma Exchange Steroids  Rituximab | Improved |
| Yoshida et al. | Japan  (2022) | 57  Male | Acute hepatitis  of unknown cause | First | Fatigue  Loss of appetite  Jaundice | Pfizer-BioNTech  First dose  7 days | Hemoglobin, 5.5 g/dl  Retic, 496\* 109/L  Platelets, 9\*109/L  Schistocytes, 17.6%  Creatinine, 138.87 µmol/L  LDH, 2275 UI/l  Haptoglobin, 3 mg/dl | <0.5%  1.9 BU/ml | Plasma Exchange Steroids  Rituximab | Improved |
| Picod et al. | France (2022) | 36  Female | Systemic lupus erythematosus | First | Bruising  Headache | Pfizer-BioNTech  First dose  6 days | Hemoglobin, 10g/dl  Platelets, 10\*109/L  Schistocytes, 3%  Creatinine, 86.24 µmol/L | <5%  0.5 BU/ml | Plasma Exchange Steroids  Rituximab | Improved |
| 54  Male | TTP | Relapse | Bruising  Diffuse  mucocutaneous  bleeding  Headache  Amnesia | Moderna Biotech/ First dose/ First dose/ 23 days | Hemoglobin, 11.5 g/dl  Platelets, 17\*109/L  Schistocytes, 2 %  Creatinine, 149.6 µmol/L | <5%  1.1 BU/ml | Plasma Exchange Steroids  Rituximab Caplacizumab | Improved |
| 60  Female | TTP | Relapse | Cerebellar  Syndrome | Pfizer-BioNTech  First dose  10 days | Hemoglobin, 10.8g/dl  Platelets, 27\*109/L  Schistocytes, 2%  Creatinine, 66.88 µmol/L | <10%  Positive | Plasma Exchange Steroids  Rituximab | Improved |
| 60  Female | No | First | Cerebellar  Syndrome  Aphasia  Confusion  Chest pain | Pfizer-BioNTech  First dose  12 days | Hemoglobin, 6.5g/dl  Platelets, 20\*109/L  Schistocytes, 6%  Creatinine, 80.96 µmol/L | 5%  52 U/ml | Plasma Exchange Steroids  Caplacizumab | Improved |
| 38  Male | No | First | Fever  Headache  Hemiparesis  Bruising | Pfizer-BioNTech  Second dose  30 days | Hemoglobin, 6.6g/dl  Platelets, 9\*109/L  Schistocytes, 5 %  Creatinine, 88.88 µmol/L | <1%  Positive | Plasma Exchange Steroids  Rituximab Caplacizumab | Improved |
| 68  Male | Mixed connective tissue disease  TTP | Relapse | Dizziness | Pfizer-BioNTech  First dose  17 days | Hemoglobin, 10.9g/dl  Platelets, 39\*109/L  Schistocytes, 1 %  Creatinine, 69.52 µmol/L | 2%  - | Plasma Exchange Steroids  Rituximab Caplacizumab | Improved |
| 66  Male | No | First | Facial paralysis | AstraZeneca-Oxford  First dose  8 days | Hemoglobin, 7.9 g/dl  Platelets, 11\*109/L  Schistocytes, 4 %  Creatinine, 81.84 µmol/L | <5%  - | Plasma Exchange  Steroids  Rituximab  Caplacizumab | Improved |
| 70  Female | Ischemic strokes  Hypertension | First | Coma  Hemiparesis | AstraZeneca-Oxford  First dose  10 days | Hemoglobin, 8 g/dl  Platelets, 6\*109/L  Schistocytes, 2%  Creatinine, 79.2 µmol/L | 11%  140 U/ml | Intravenous Immunoglobulins Plasma Infusion Steroids  Rituximab Caplacizumab | Death 2 month  after presentation |
| 22  Male | No | First | Coma  Seizures  Purpura  Fever | Pfizer-BioNTech  Second dose  18 days | Hemoglobin, 6.8g/dl  Platelets, 10\*109/L  Schistocytes, 2%  Creatinine, 101.2 µmol/L | 6%  Positive | Plasma Exchange Steroids  Rituximab | Improved |
| 20  Female | Systemic lupus erythematosus | First | Systemic lupus erythematosus Flare  Polyarthritis  Erythema | Pfizer-BioNTech  First dose  25 days | Hemoglobin, 5.3 g/dl  Platelets, 51\*109/L  Schistocytes, 3%  Creatinine, 88 µmol/L | <10%  50 U/ml | Plasma Infusion Steroids | Improved |

\*Autoantibodies to ADAMTS-13 was assessed either as the titer of total autoantibodies with a simplified enzyme-linked immunosorbent assay (ELISA) and expressed in arbitrary units (U/mL; normal < 12 U/mL) or as the titer of inhibitory antibodies using an alternative methodology (Bethesda assay) expressed in Bethesda Units (BU/mL; normal <1 BU/mL) or BEU (normal <0.4). NA: Not available. Retic, reticulocytes; LDH, Lactate dehydrogenase.