**Suppl 1**. Literature Review

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|  | **Study** | **Number of cases** | **Median age** | **M/F** |  **Associated malignancy B-cell lymphoma (n)/T-/NK-cell lymphoma (n)** |  **Fever** | **Hepatospleenomegaly** | **Cytopenias** | **Hypertriglyyceridemia** | **Hyperferretinemia** | **Hypofibrinogen** | **NK-cell activity/soluble CD25/IL-2** | **Hemophagocytosis** | **EBV ISH +** | **NHL/HLH therapy**  | **Outcome/status** | **Median survival from HLH diagnosis** |
| 1 | Han et al, 2007 [10] | 29 | 46 ( 9 - 87) | 16:13 | 5/24 | 29 (100%) | 27 (93.1%) | 25 (86.2%) | NA | 13 (92.9%) | 15 (57.7%) | NA | NA | 13 (68.4%) | 23 | CR (4), PR (3) | 36 days |
| 2 | Jiang et al, 2016 [6] | 41 | 44 (24.5 - 54.0) | 28:13 | 29/12 | 41 (100%) | 36 (87.8%) | NA | NA | NA | NA | 40 (97.5%) | 32 (78%) | NA | NA |  Dead (26) | 120 days |
| 3 | Li et al, 2014 [11] | 16 | 48.5 (15 - 70) | 10:6 | 1/15 | 16 (100%) | 13 (81.3%) | 16 (100%) | 10 (62.5%) | 16 (100%) | 15 (93.8%) | NA | 9 (56.3%) | 7 (43.8%) | 13 (81%) | CR (1), PR (2), dead (15) | 37 days |
| 4 | Yu et al, 2013 [8] | 30 | 51 (21 - 77) | 13:17 | 13/17 | NA | 26 (86%) | 24 (80%) | NA | NA | NA | NA | 20 (66%) | 9 (30%) | 30 (100%) | CR (9), PR (2), dead (23)  |  231 days |
| 5 | Cattaneo et al, 2016 [9] | 24 | NA | NA | 10/14 | NA | NA | NA | NA | 24 (100%) | NA | NA | NA | 10 (41%) | 24 (100%) | CR (9) | 87 days |
| 6 | Parikh et al, 2014 [7] | 32 | 54 (18 - 87) | 22:10 | 9/19  | NA | NA | NA | 9 (43%) | 25 (100%) | 30 (55%) | NA | NA | NA | 25 (84%) | NA | 42 days |

NA: not available; CR: complete remission; PR: partial remission.

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