**Supplemental Table 1: Synopsis of treatments for protein-losing enteropathy.**

|  |  |
| --- | --- |
|  |  Number of patients |
| **Medical treatment** | Total | Remission | Relapse |
| Budesonid + sildenafil  | 8 | 4 | 4 |
| Budesonid + sildenafil + bosentan | 5 | 3 | 3 |
| Budesonid + sildenafil + prostacyclin | 1 | 0 | 1 |
| Budesonid | 2 | 1 | 1 |
| Sildenafil  | 5 | 4 | 1 |
| Sildenafil + bosentan | 1 | 0 | 1 |
| Prostacyclin | 1 | 0 | 1 |
| Heparin | 7 | 3 | 4 |
| **Surgical treatment** |  |  |  |
| Diaphragm plication | 11 | 7 | 4 |
| AV valve reconstruction / replacement | 2 | 1 | 1 |
| Pacemaker implantation | 5 | 1 | 4 |
| Creation of fenestration | 2 | 1 | 1 |
| Fontan revision | 2 | 0 | 2 |
| Other | 3 | 1 | 2 |
| **Interventional treatment** |  |  |  |
| Balloon angioplasty / stent implantationpulmonary arteries | 14 | 9 | 5 |
| Balloon angioplasty / stent implantation IVC / SVC | 8 | 3 | 5 |
| Embolization of collaterals | 16 | 7 | 9 |
| EPS | 1 | 0 | 1 |
| Creation / dilatation of fenestration | 3 | 0 | 3 |
| Other | 6 | 2 | 4 |
|  |  |  |  |

This tabulated synopsis summarizes the treatment strategies performed during follow-up in patients with protein-losing enteropathy relapses which are depicted in in Fig. 2. Treatments are summarized for the entire cohort analyzed as well as for the subgroups of patients with remission and those without remission or frequent relapses; data are presented as absolute figures. Heparin treatment was partially overlapping with other medical treatments and also medical treatments were combined with interventional and surgical treatments in various patients (Fig. 3).

AV valve - atrioventricular valve; ICV - inferior vena cava; SVC - superior vena cava, EPS - electrophysiological study.